An Atlas of Esophageal Motility

In Health and Disease

AN ATLAS OF

ESOPHAGEAL MOTILITY

in Health and Disease

This Atlas is from the Section of Physiology and the Section of Medicine, Mayo Clinic and Mayo Foundation. The Mayo Foundation, Rochester, Minnesota, is a part of the Graduate School of the University of Minnesota.

by

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Introduction

Studies of the motor activity of the human esophagus have been in progress in our laboratory during the past 7 years. Modern recording devices have provided simple, clearly defined, pictorial representations of complex sequences. Publication in scientific journals has allowed presentation of only a few of these tracings. Yet, as experience accumulated, we became convinced that the records tell the story themselves—more definitively and more quickly, indeed, than the spoken or written word. Once satisfied of this, we determined to set forth our findings in atlas form. This volume is the result. Its purpose is to present pictorially the motor activity of the human esophagus in healthy persons and in patients with certain diseases. The illustrative material is limited to that collected in our own laboratory. Excellent recordings of events in the esophagus have been published by others, but since we did not have first-hand acquaintance with these, they have not been reproduced. To this extent the presentation is biased. However, we offer no apology, since it was our intent from the onset that the volume rest on experience accumulated in our laboratory.

An effort has been made to keep the written word to a minimum, to let the illustrations tell their story. Reference to the literature has been made in the text mainly to acknowledge observations of others which interlock with our own, and to guide the scholar to more detailed sources of information. The bibliographic material is, however, limited, and wider sources should be sought if review is contemplated.

Acknowledgments

Much of the experience which this volume illuminates was acquired while we worked with younger colleagues. They struggled with details of technic until satisfactory procedures were evolved, and they gathered recordings from healthy persons and patients with certain diseases until sufficient numbers were available to make generalizations possible. Those who have made such contributions without aiding directly in the preparation of this volume are Nicholas C. Hightower, Jr., J. Walker Butin and F. Earl Fyke, Jr. We, the authors of this volume, wish to express our sincere thanks to each. Without their earlier participation this volume would not have been possible. We are also very grateful to Alan W. M. Smith, who aided in the organization of the material used in this volume.

Once we had selected the illustrative material for this atlas, it was necessary to mount and to label each new tracing and to reassess previously prepared material for this new use. Many new illustrations had to be made and much photographic material processed and reprocessed. We are grateful to Mrs. Jean Frank and to Mr. Stanley McComb for their patience with us, their recommendations to us, their meticulous attention to detail and their willingness at all times to do and re-do any illustration if improvement was in prospect.

The Authors

CONTENTS

An Atlas of Esophageal Motility In Health and Disease

CHAPTER ONE

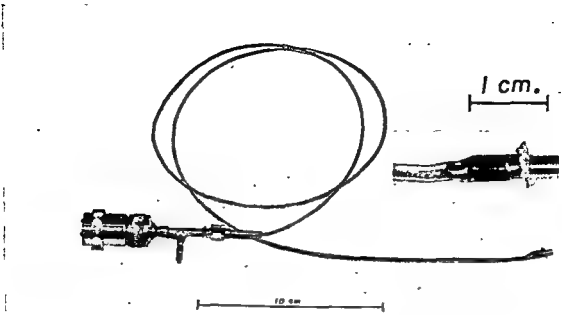
Methods and Procedures

Motor activity in the esophagus and its sphincters was determined by measurement of intraluminal pressures. This was accomplished by the use of tiny electromagnetic pressure transducers. In some tests, a single transducer was passed into the esophagus. In others, water-filled tubes were swallowed by the subjects or patients and these were then connected to transducers situated just outside the mouths of the subjects or patients. The pressures detected by these devices are those occurring within the lumen of the esophagus in the immediate vicinity of the unit. At times, with the esophagus relaxed and patent, the change in pressure may be common to the entire length of the organ. In other instances, when the walls of the esophagus or its sphincters are in direct contact with the pick-up unit, the changes in pressure which it detects are localized. For example, circular contraction of the walls of the esophagus may squeeze directly against the pressure-sensitive orifices of the units or against tiny pockets of air or fluid surrounding them.

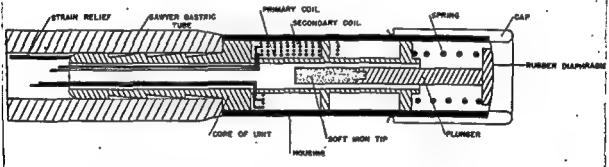
The miniature electromagnetic pressure transducer is housed within a capsule which is 3 mm. in diameter and 9 mm. in length. It is attached to the end of an esophageal tube (upper panel).

The principle of the transducer is that of the transformer or the induction coil. The present model is based on the modification by Gauer and Gienapp[1, 2] of the original design by Wetterer.[3, 4] The intraluminal pressure moves a plunger, the tip of which is soft iron and lies within the coils of a differential transformer (center panel). Movement of the iron tip creates an imbalance in the current induced in the two sets of secondary coils. This signal, after suitable amplification, activates a galvanometer.

In spite of the small size of the instrument, the frequency response of the transducer is within 5 per cent of the static sensitivity up to 10 cycles per second.

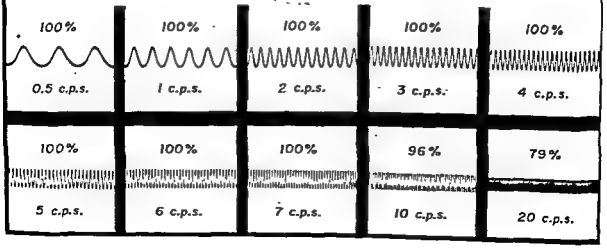


SCHEMATIC DIAGRAM OF MINIATURE PRESSURE TRANSDUCER



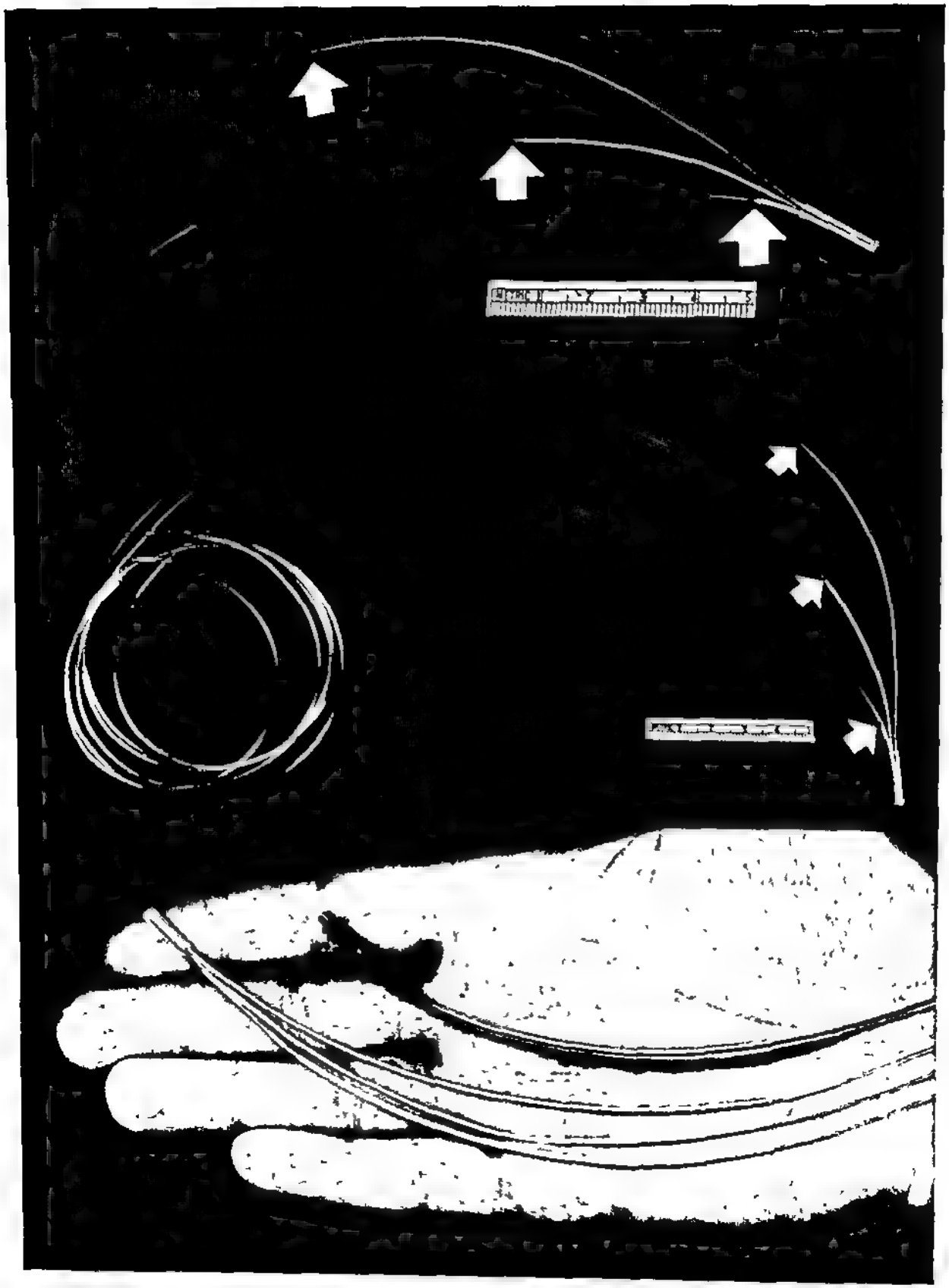
DYNAMIC RESPONSE OF TRANSDUCER SYSTEM

Per Cent Static Sensitivity At Various Frequencies



Because of its size, one transducer is all most people can swallow with comfort. Simultaneous recordings of the changes in pressure from multiple sites are made by the use of polyethylene tubes to conduct the pressures to transducers located outside the body. Three tubes with an inner diameter of about 0.03 inch and an outer diameter of about 0.05 inch are fastened together at the tips, and a lateral hole is cut in each. The holes usually are separated by 5 cm. Since these tubes with lateral holes transmit changes in pressure much as tubes with open tips do, they have been referred to, for convenience, as "open-tip tubes."[7] The distal part of each tube, beyond the orifice, is filled with mercury, which enables the exact level of the orifice to be determined roentgenographically.

The three open-tip tubes are less bulky than a single transducer.



ch open-tip tube is connected to a transducer through a four-way stop-
The tubes are filled with water from syringes attached to the stopcocks.
ring a test are kept patent by intermittent flushing with water.

andard pressures are introduced into the system at the beginning and
end of each test.

During an actual test, errors referable to the hydrostatic pressure exerted by the column of water in the tubes are minimized by the placing of the transducers, stopcocks and flushing syringes at the same level as that of the orifices of the tubes.

METHODS AND PROCEDURES

The pressure pick-up unit is passed through the mouth and advanced until the tip is in the fundus of the stomach. The myograph and pneumograph are attached and the subject then rests in a supine position on an x-ray table. A continuous recording of resting pressures is made as the detecting unit is withdrawn at intervals of 0.5 cm. through the gastroesophageal junction. The single unit is then returned to the stomach and the sphincteric zone is re-explored as the subject swallows after each withdrawal of 0.5 cm. When three open-tip tubes are used, the resting pressures usually are determined by withdrawal of the first two and the response of the zone to deglutition during withdrawal of the third at the same intervals of 0.5 cm.

The response of the esophagus to deglutition is recorded at intervals of 1 or 2 cm. throughout the entire length of the structure.

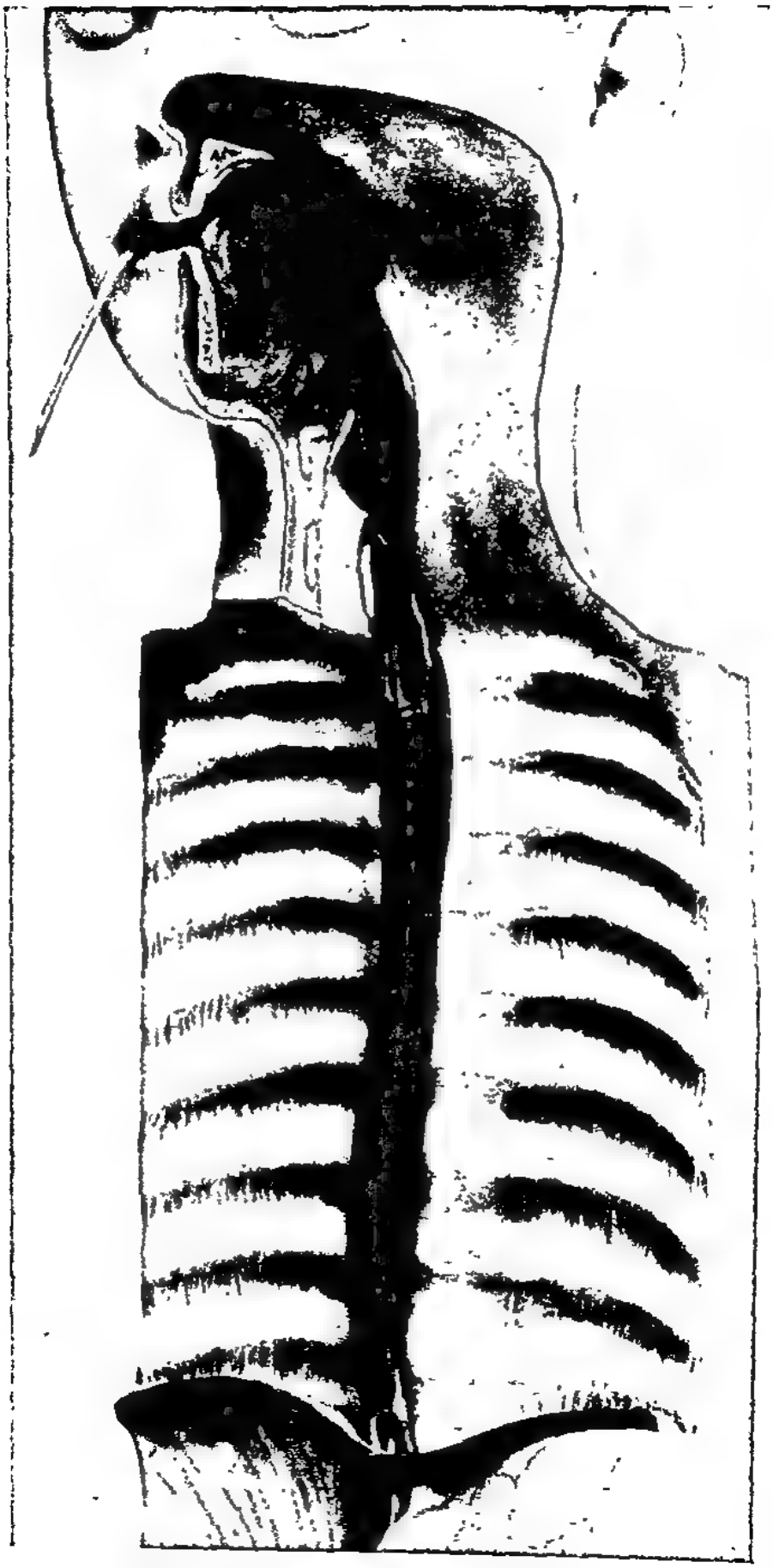
The pharyngo-esophageal junction is examined in the same manner as is the gastroesophageal junction.

The exact distance of the unit from the incisor teeth is measured at each recording site. The site is approached by withdrawing the pressure-sensitive unit into the region, rather than by advancing it.

CHAPTER TWO

The Esophagus and Its Sphincters In Health

From a physiologic point of view, the esophagus may be regarded as a muscular tube closed at both ends by sphincters. In health, the tube and its sphincters operate as a functional unit which responds in a co-ordinated manner to deglutition or esophageal distention. Between such responses, the variations in pressure within the esophagus are reflections of changes occurring outside the organ. A relatively constant or steady state exists in the walls. This has been taken to represent the "resting" condition of the esophagus.



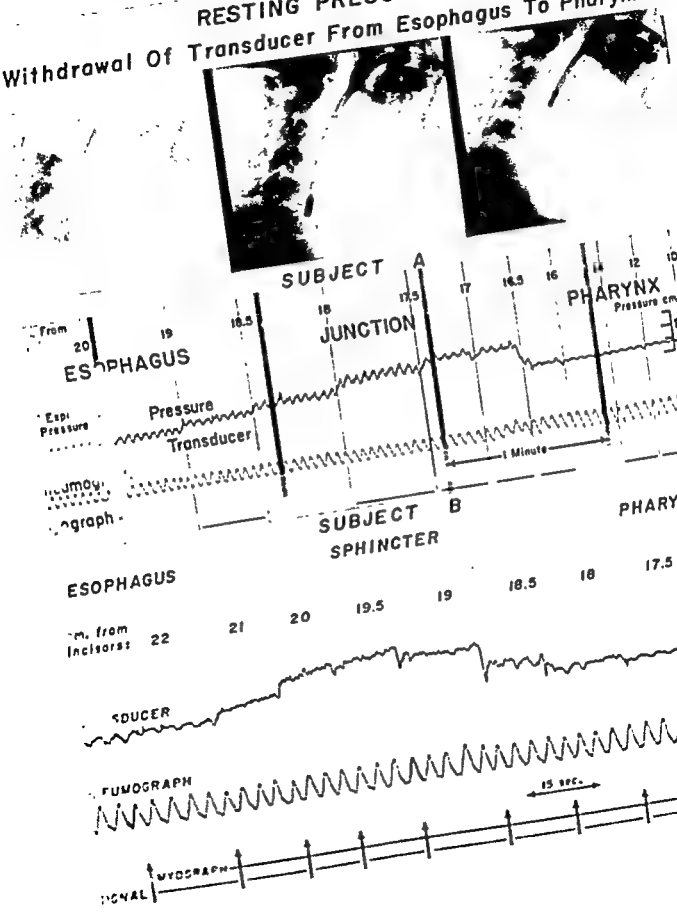
Pharyngo-esophageal Junction

HEALTH

Pharyngo-esophageal Junction

RESTING PRESSURES

Withdrawal Of Transducer From Esophagus To Pharynx

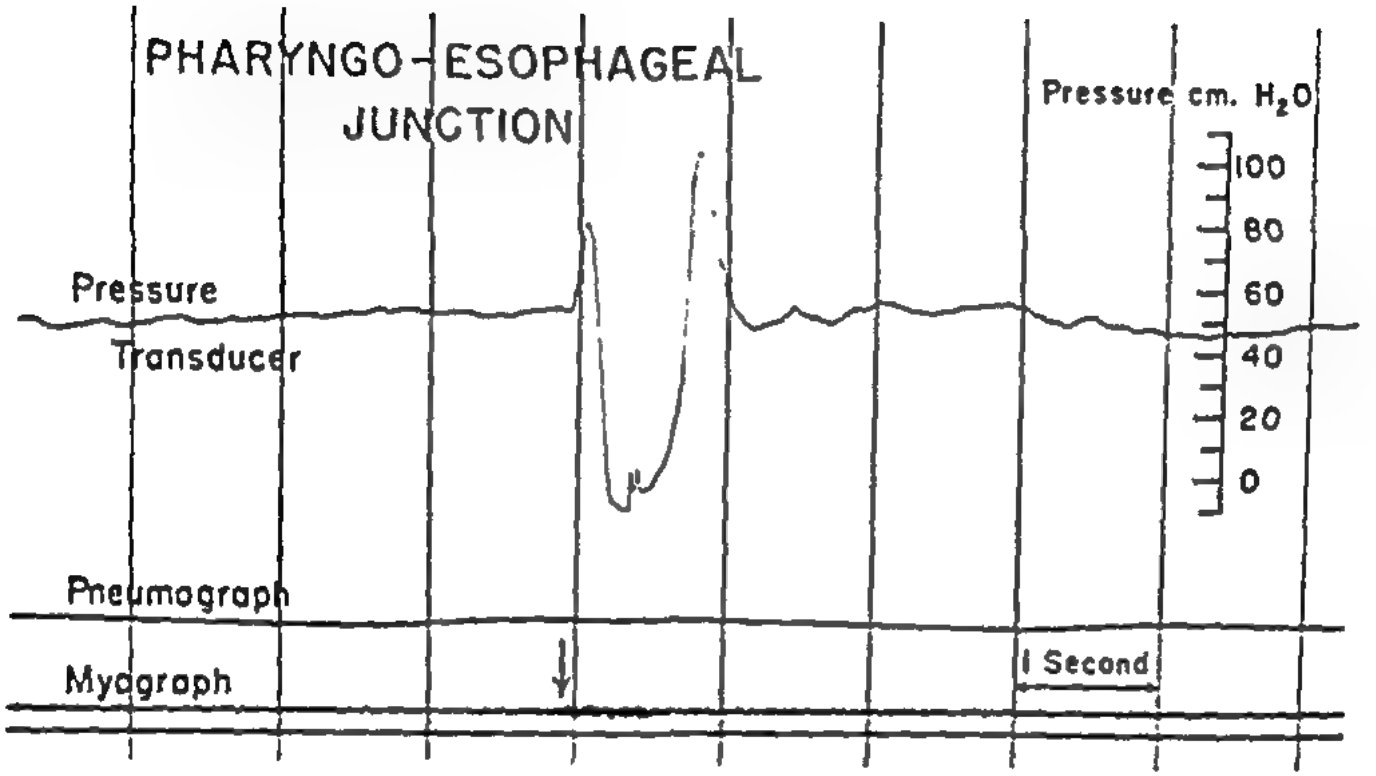


Resting Pressures

In the resting state, the pharyngo-esophageal sphincter is represented by a zone of elevated pressure 2 to 3 cm. in width, lying between the upper part of the esophagus and the pharynx. This area can be clearly delineated by withdrawal of a pressure-sensitive device from the upper part of the esophagus into the pharynx. As the withdrawal progresses, the pressure increases until, in the upper portion of the zone, it is 20 to 30 cm. of water above the pressure in the upper part of the esophagus. When the device is withdrawn farther, the pressure decreases abruptly as the unit enters the pharynx.[8]

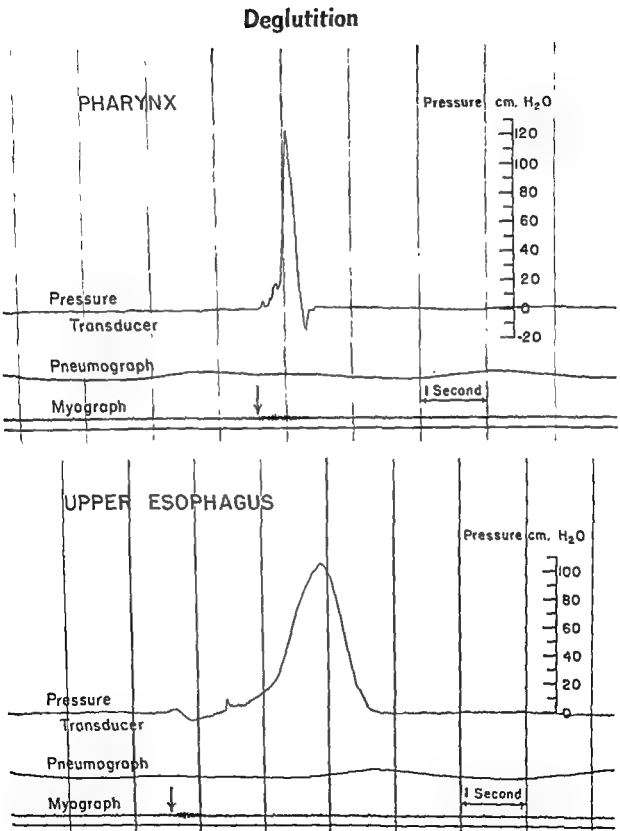
The band of elevated pressure is an effective barrier between the pharynx and the esophagus.

Deglutition



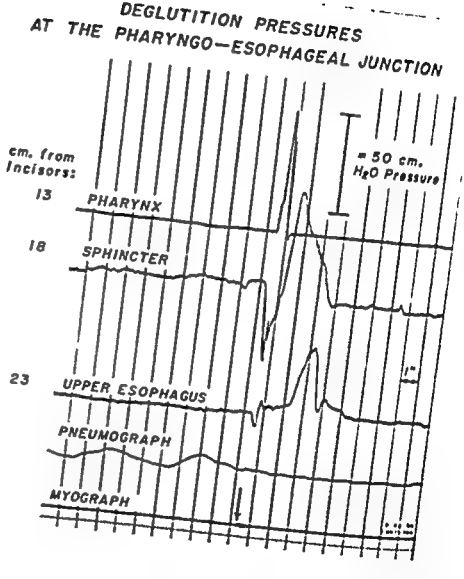
When the miniature transducer lies in the high-pressure zone of the meter, swallowing causes a decrease in pressure, indicating relaxation of the meter.

In these and subsequent records, the arrow above the myographic recording cts attention to the burst of action potentials from the voluntary muscles of deglutition, and marks the onset of the act of swallowing.

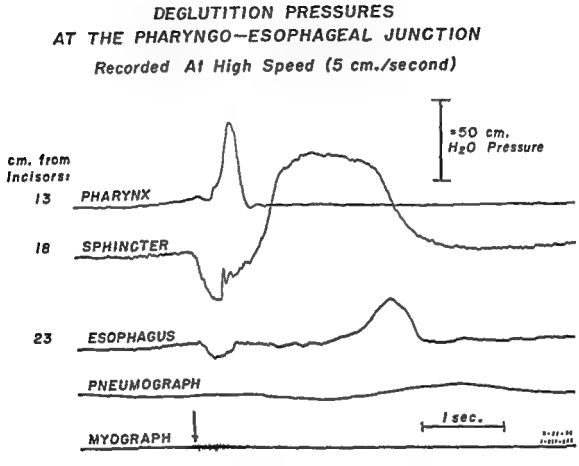


While the sphincter relaxes, the pharynx contracts and this contraction passes *later into the esophagus.*[9]

Deglutition

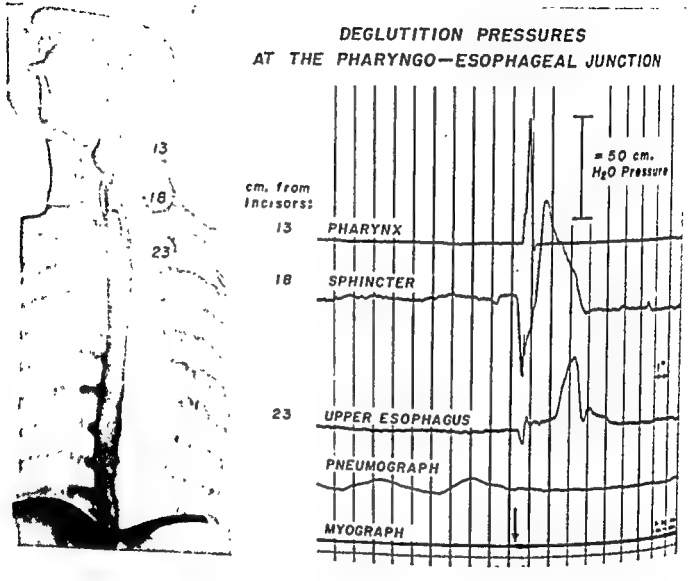


Simultaneous recordings from three sites clearly depict the sequence. The vated resting pressure in the sphincter is abolished as the pressure in the rynx is increased. The increase of pressure then passes as a wave through the sphincter into the upper part of the esophagus.



The intimate temporal details of the sequence are more clearly depicted when a high-speed recording is made. It is clear, then, that the sphincter relaxes just prior to the occurrence of the high pressure in the pharynx, thus ensuring that the doorway to the esophagus is open as the swallowed material is forced toward it by the pharyngeal contraction. The contraction then passes in a co-ordinated manner through the sphincter into the upper part of the esophagus. The prolonged contraction of the sphincter during the peristaltic sequence is characteristic of the sphincteric zones of the esophagus.

Deglutition



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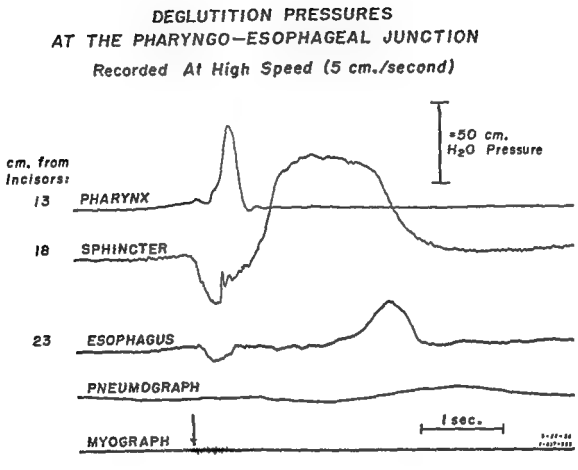

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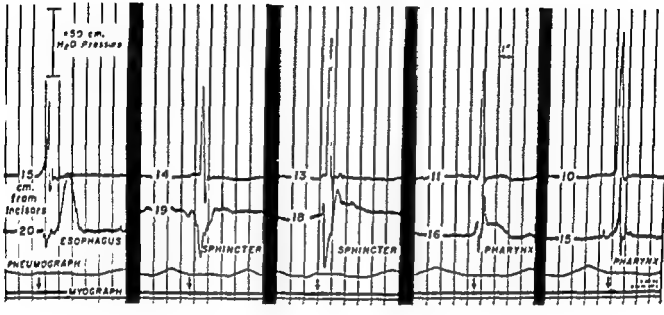
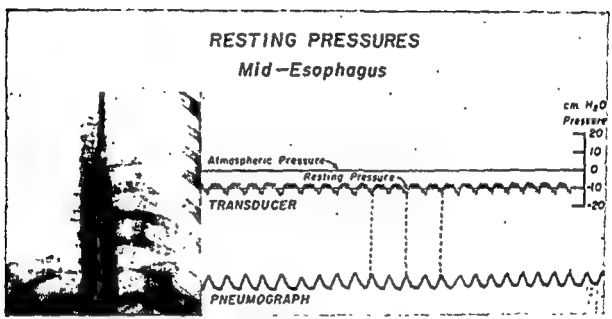


The participation of the zone in the sequence of deglutition is clearly evident when one of two detecting units is withdrawn at intervals of 1 cm. through the sphincter. In the left-hand panel, the two units straddled the sphincter. In the middle panel the lower unit was in the center of the sphincteric band, while in the final panel, both units were in the pharynx.

Pressures in the Body of the Esophagus

Resting Pressures



Resting pressures in the esophagus are subatmospheric, and the major fluctuations are due to breathing. With inspiration the pressure becomes more negative.[10]

In this and all other records, an upward deflection of the pneumographic tracing indicates inspiration, and a downward deflection denotes expiration. *End-inspiration then occurs at the apex of each respiratory swing of the pneumograph*, while end-expiration is marked by the base of the swing.

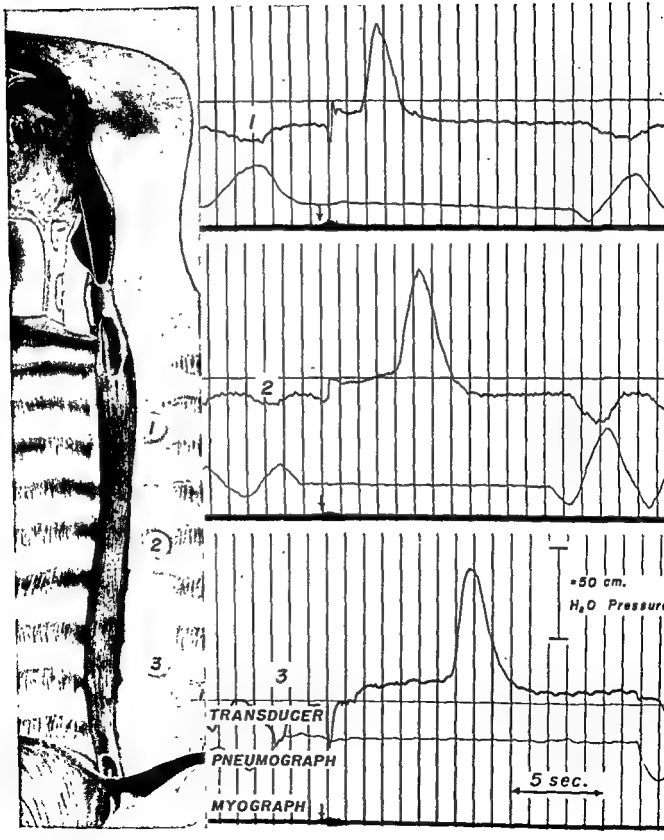
Pressures in the Body of the Esophagus

Deglutition Pressures

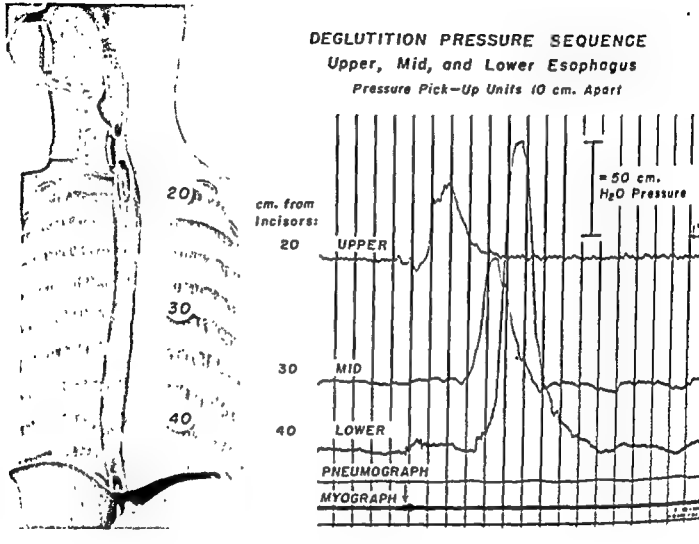
Deglutition

Swallowing produces a complex increase of pressure in the esophagus, the details of which are recorded most clearly when the subjects swallow while the breath is held. A single pressure pick-up unit may be used to detect the details of the changes in pressure at different levels in the esophagus.[1] As swallowing occurs, a slight negative deflection is often seen in the upper and lower parts of the esophagus (1 and 3). When a sip of water is swallowed, the pressure suddenly increases as the water is forced into the esophagus by the pharyngeal contraction (1, 2 and 3). Contraction of the esophagus produces the most prominent pressure peak in the complex. This high pressure sweeps in sequence from the upper, through the middle and into the lower part of the esophagus, reaching the depths of the organ about 9 seconds after swallowing (3).

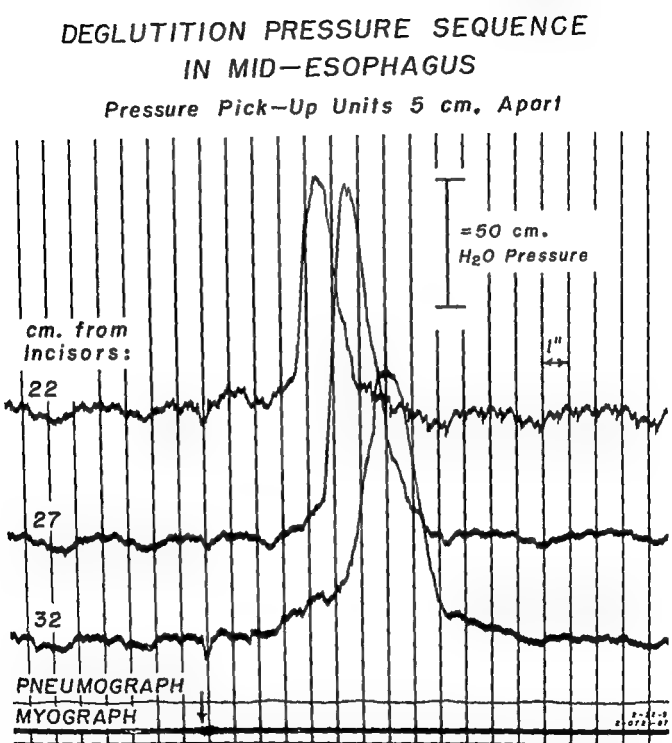
Deglutition



Deglutition

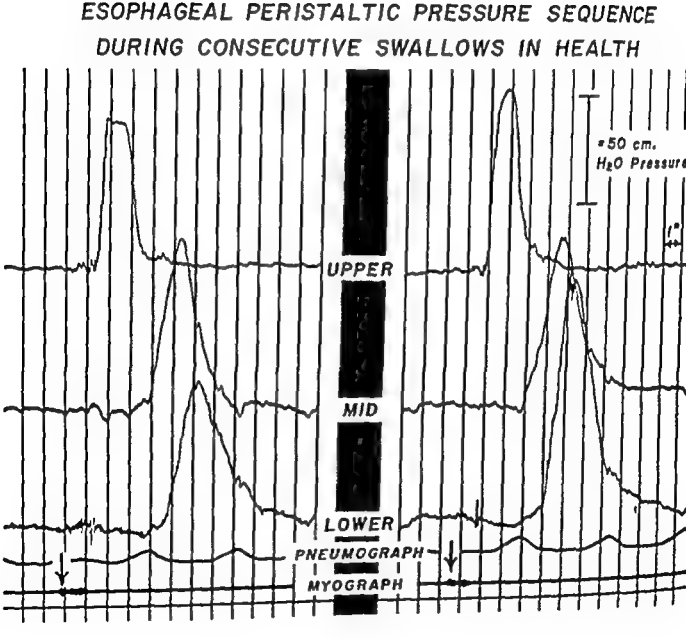


The sequential characteristics of the main changes in pressure in the esophagus after the act of swallowing are most satisfactorily delineated by simultaneous recordings from multiple sites. The increase in pressure is detected first in the upper part of the esophagus and last in the lower part of the esophagus.



The contraction of the esophagus in response to swallowing passes as a peristaltic wave over the length of the organ, and not as a series of segmental contractions. Shortening the distance between the pick-up units demonstrates the uninterrupted progression of the contraction.

Deglutition



Deglutition

Consecutive swallows produce very similar pressure sequences in the esophagus of a healthy person.

The Gastroesophageal Sphincter

Gastroesophageal Sphincter

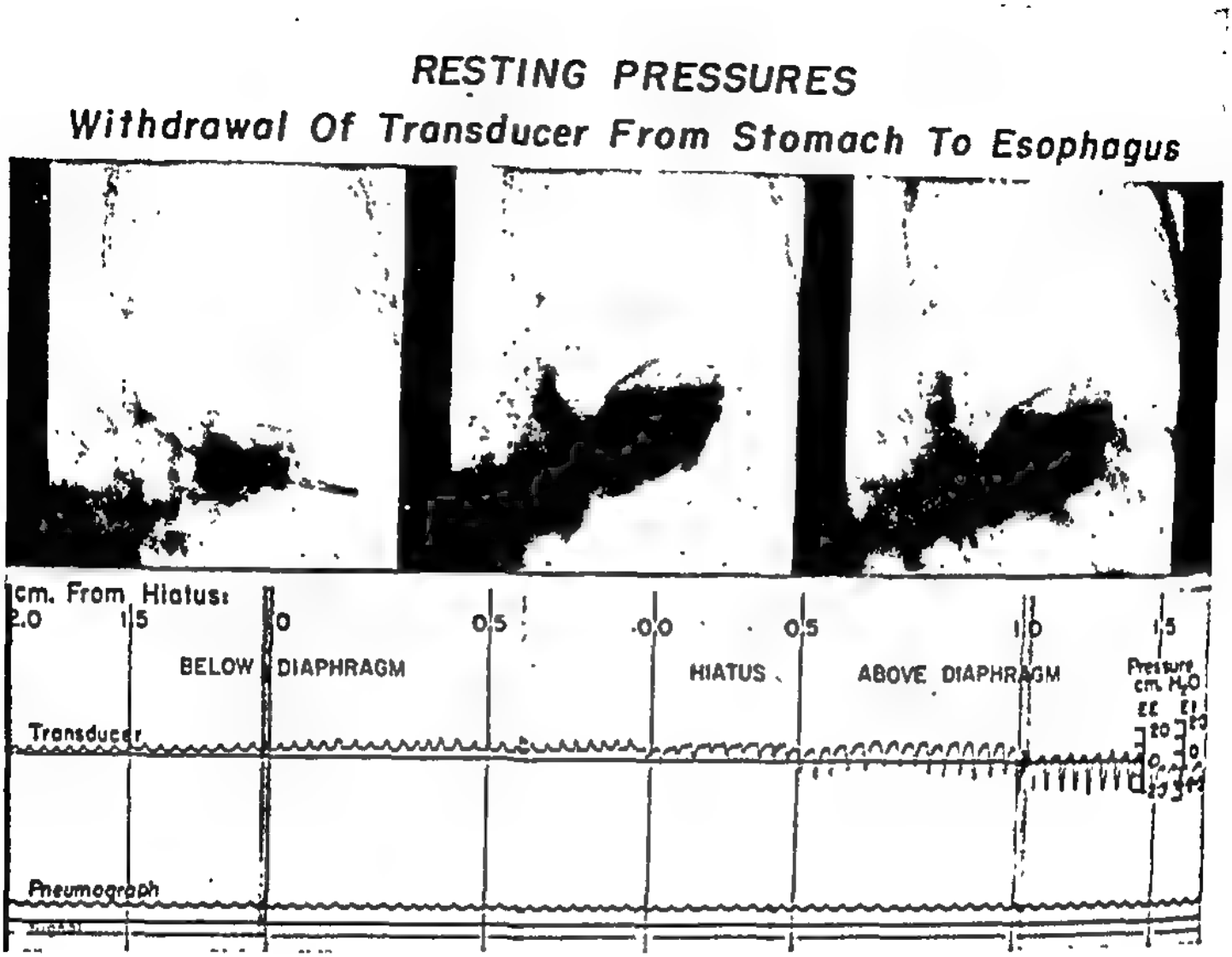
In health, a band or zone of elevated pressure is interposed between the stomach and the esophagus. In the central region of this zone, the pressure swings with breathing become reversed. With deglutition or esophageal distension, this high-pressure band is temporarily abolished.[12]

Resting Pressures

As a pressure-sensitive device is withdrawn from the stomach into the esophagus, the direction and amplitude of the fluctuations in pressure with breathing change. In the stomach the pressure increases with inspiration, while in the esophagus it decreases. This reversal of the pressures with breathing occurs at the diaphragm. Within the esophagus, the transition from subdiaphragmatic to supradiaphragmatic patterns usually is complete as the unit is withdrawn through only 0.5 cm. of the zone. Often the change is abrupt. Sometimes the unit may come to rest at a point within the zone at which no change of pressure with breathing is detected, and occasionally biphasic waves are recorded which reflect both subdiaphragmatic and supradiaphragmatic types of pressure swings with each breath. Whatever the pattern, the point of respiratory reversal always can be recognized. This point thus affords a convenient physiologic landmark above and below which distances can be measured accurately. In health, this point lies at, or in close association with, the esophageal hiatus of the diaphragm, and has therefore been labeled "hiatus."

Resting Pressures

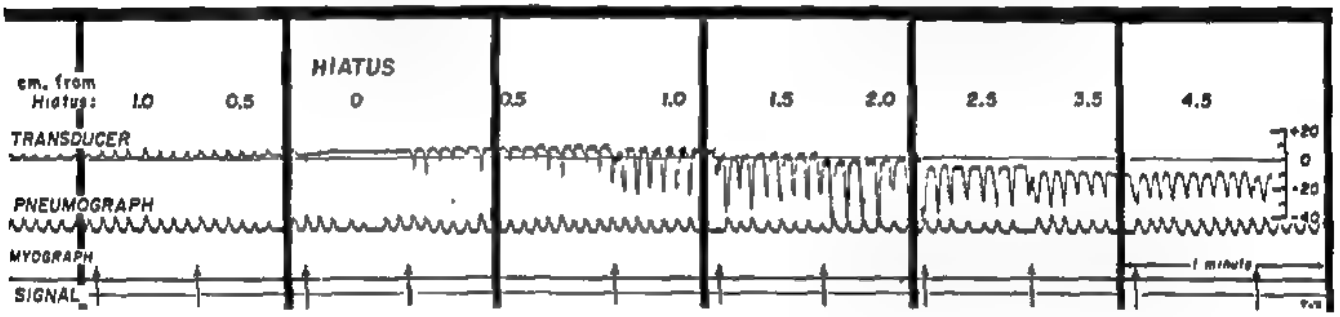
In health, the resting pressures in the stomach are always in excess of those in the esophagus. If there were no barrier between the two organs, material would flow from the stomach to the esophagus. However, during inspiration, the pressure over a band about 1.5 cm. wide, just below the point of respiratory reversal, is always greater than the pressure within the stomach. During expiration, the pressure in this area is slightly increased over that in the stomach, but for a distance of about 2 cm. just above the point of respiratory reversal it is higher than that in the stomach. Thus a pressure barrier is always interposed between the stomach and the esophagus. This barrier or band of elevated pressure is regarded as due to the tonic contraction of circularly disposed muscle fibers.



In this test the single pressure transducer was withdrawn at intervals of 0.5 cm. from the stomach to the esophagus. Each withdrawal is indicated by a thin vertical line drawn through the break in the signal. The thicker gray lines delineate minutes. The direction of the pressure swings with breathing changed abruptly at the withdrawal labeled 0.0. A slight increase in pressure commenced about 1.5 cm. below, and continued for about 1 cm. above, this point. The zeros on the calibrations are the resting pressures in the fundus of the stomach at end-expiration (EE) and at end inspiration (EI). A line has been drawn to indicate the level of end-expiratory fundic pressure.

Resting Pressures

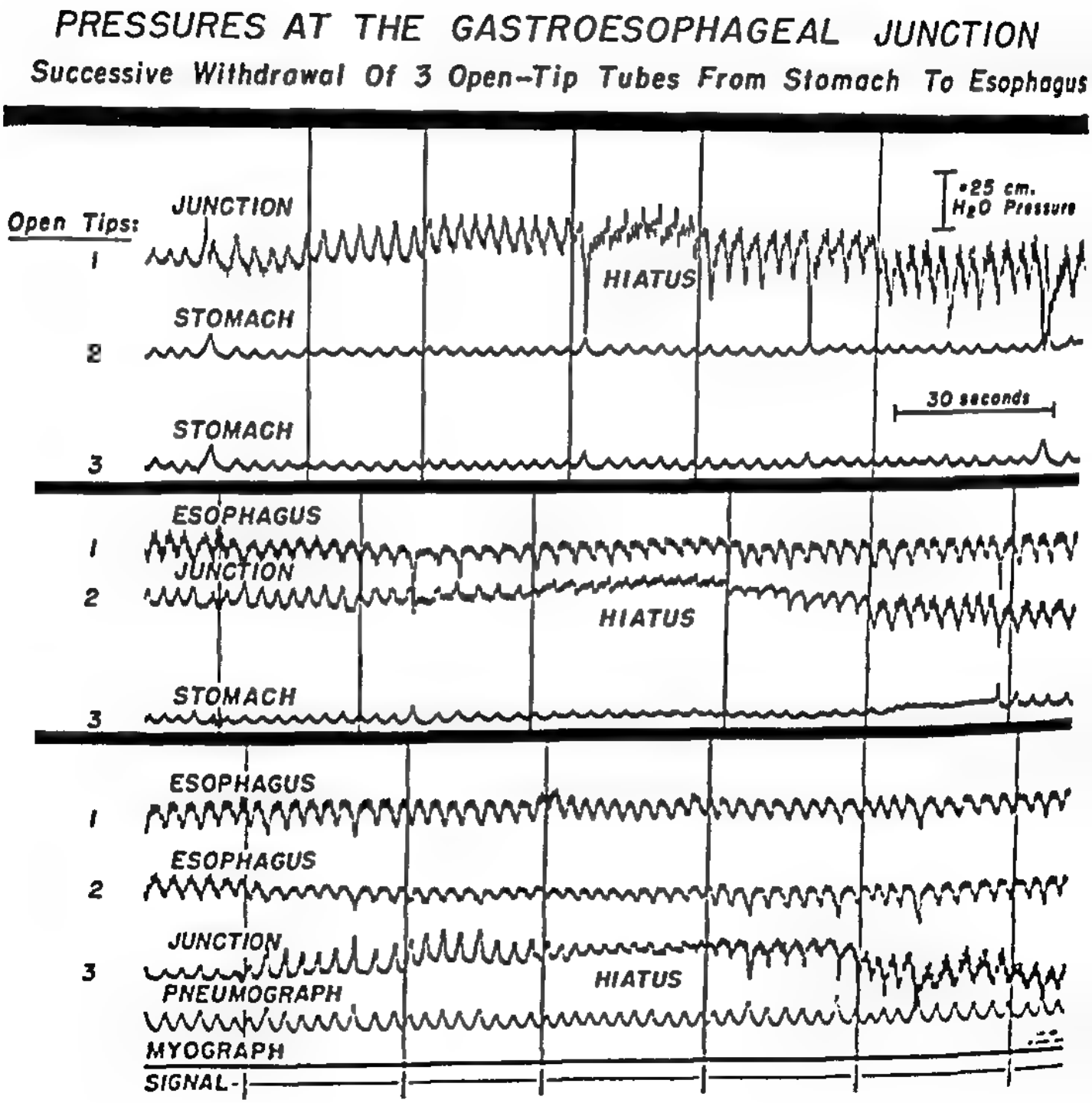
RESTING PRESSURES AT THE GASTROESOPHAGEAL JUNCTION IN HEALTH
Withdrawal Of Transducer From Stomach To Esophagus



Here the "hiatus" is identified by a zone in which no change of pressure occurred with breathing. Withdrawals are indicated by the upright arrows.

Pressures in the junctional zone are more than those in the fundus of the stomach and in the esophagus.

Resting Pressures



Resting Pressures

The three panels are parts of a continuous recording. As the three open-tip tubes passed successively through the junctional zone, each recorded an increase in pressure.

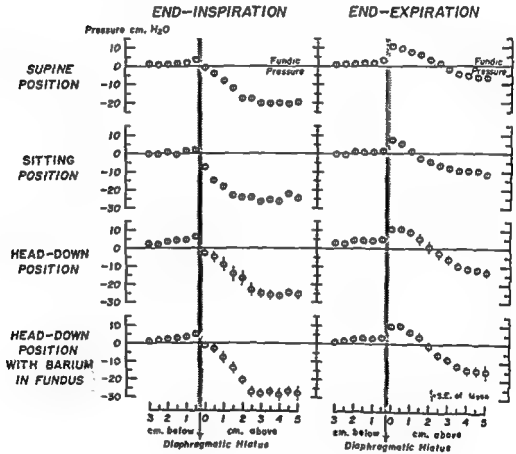
The band of increased pressure between the stomach and the esophagus represents the gastroesophageal sphincter.

Resting Pressures

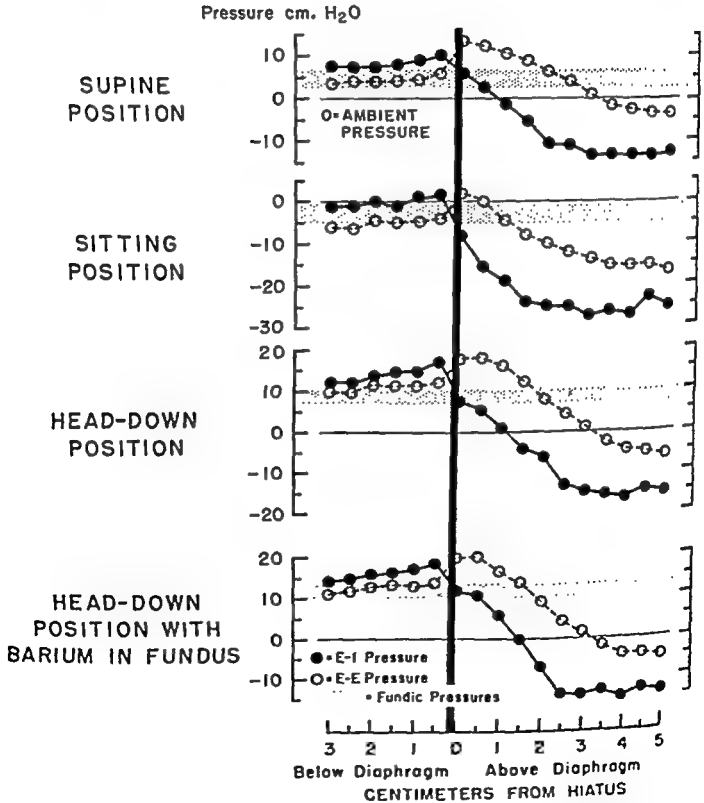
The pressure in the fundus of the stomach has been found to be very constant in an individual during a 1-to-3 hour motility test.[12] It has therefore been used as the baseline or zero pressure to which to relate the pressures encountered in the sphincteric zone. Since the pressure swings with breathing are large, those occurring at the end of inspiration and at the end of expiration have been measured.

A band of elevated pressure is always present between the stomach and esophagus, regardless of the position of the subject. At end-inspiration, however, the greatest pressures in the barrier lie below, while at end-expiration they are above the point of respiratory reversal.

MEAN RESTING PRESSURES
AT THE GASTROESOPHAGEAL JUNCTION IN HEALTH
Study Of 19 Healthy Persons



RESTING PRESSURES OF THE FUNDUS AND GASTROESOPHAGEAL REGION AND THEIR RELATION TO AMBIENT PRESSURE



Resting Pressures

The pressure barrier between the stomach and the esophagus is always retained, even though fundic pressures may be more or less than ambient, depending upon the position of the subject.

Gastroesophageal Sphincter

Deglutition

Deglutition

During swallowing, the pressure in the gastroesophageal sphincter decreases, indicating relaxation.[12] The onset of the decline occurs early in the sequence of deglutition and persists until the peristaltic wave reaches the sphincteric zone. Then, in sequence with the peristaltic contraction of the esophagus, the pressure in the sphincter increases to levels above those encountered at rest. A gradual decline to the resting values follows. The sequence indicates that the peristaltic wave enters the sphincteric zone and closes the sphincter by a contraction which is more prolonged but of less magnitude than that in the esophagus proper. This prolonged contraction of the sphincter marks the end of the act of deglutition.

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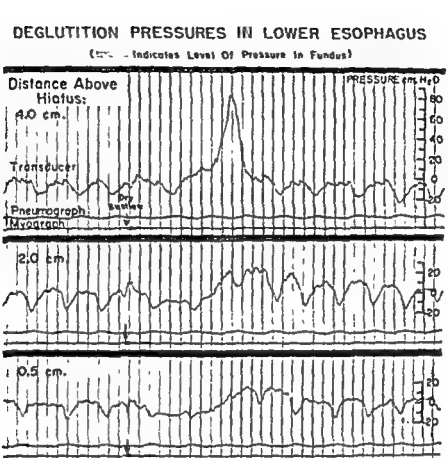
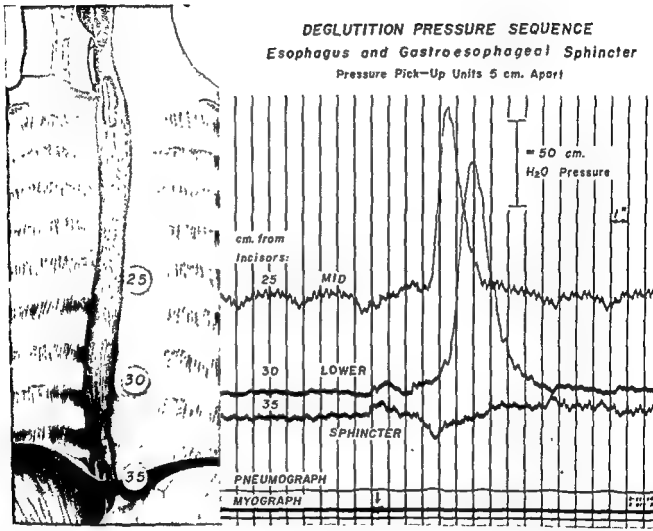


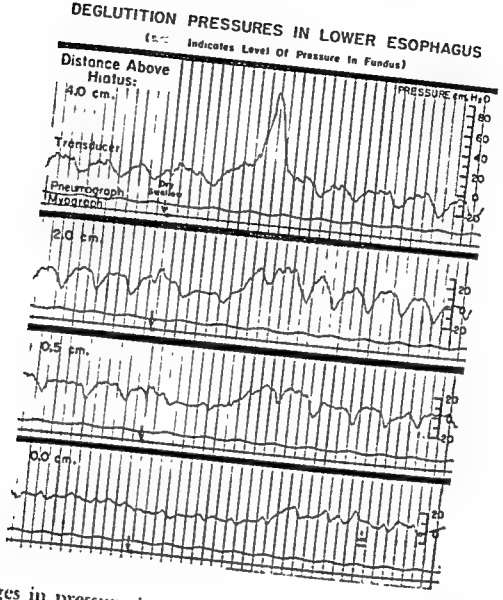
The changes in pressure in the sphincteric zone during swallowing were first detected by use of a single pressure pick-up unit. The sequence of changes through the zone was then constructed by studying the recordings made at different levels in the sphincter. Within 2 to 3 seconds of swallowing, the pressure in the sphincter declines, because of the relaxation of the sphincter.

The period of relaxation is terminated by arrival of the peristaltic wave from the esophagus. The sphincter responds with a peristaltic contraction which is of lesser magnitude, but of greater duration, than that in the lower part of the esophagus.



When multiple pick-up units are used, the temporal relationships are more apparent.

The sphincter relaxes early in the sequence of deglutition.



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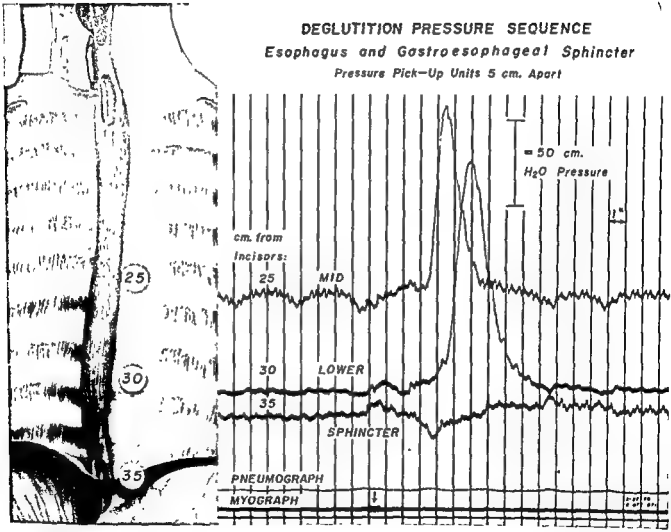

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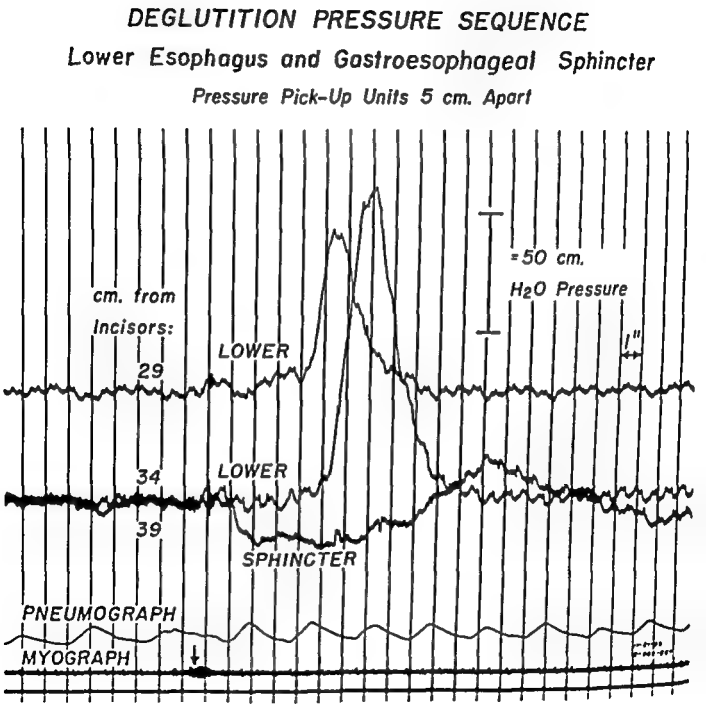
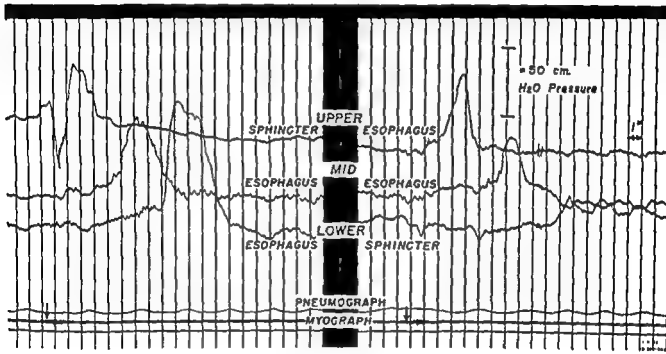


The sphincter contracts after the peristaltic wave has swept through the esophagus.

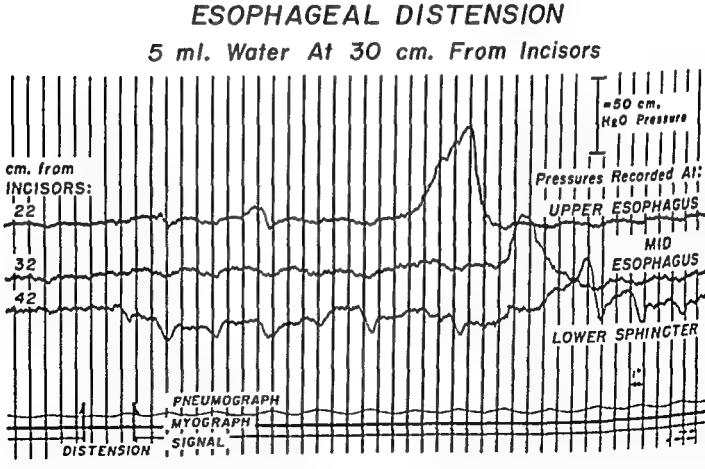


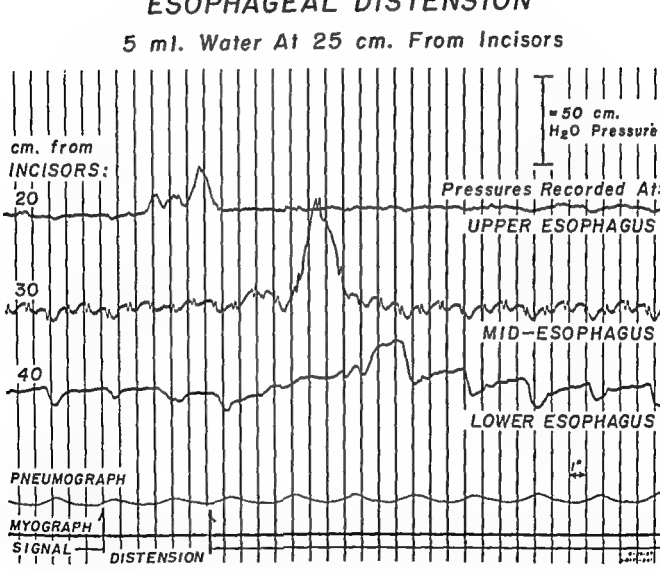
In health, the responses of the esophagus and its sphincters to deglutition are closely integrated.

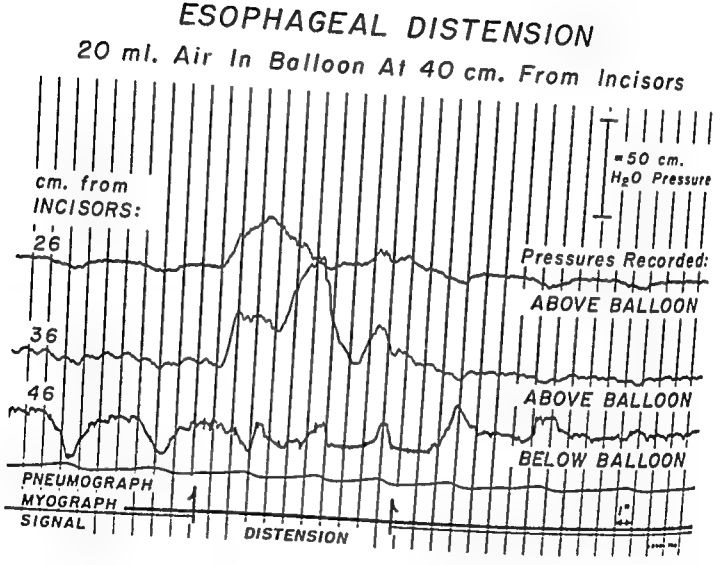
Distension of the Esophagus

Distension of the esophagus with liquid uniformly induces relaxation of the gastroesophageal sphincter and a peristaltic wave in the esophagus.[13]

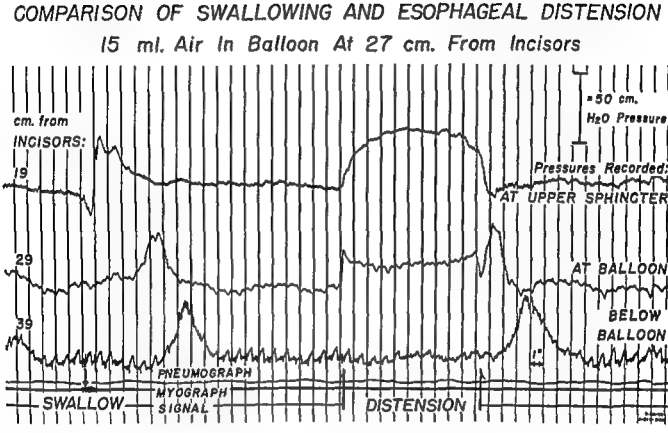
The relaxation of the sphincter occurs as the distending stimulus is applied the peristaltic wave may be initiated then, or its onset may be delayed.




ESOPHAGEAL DISTENSION
5 ml. Water At 25 cm. From Incisors
= 50 cm.
H2O Pressure
cm. from
INCISORS:
20
30
40
Pressures Recorded At:
UPPER ESOPHAGUS
MID-ESOPHAGUS
LOWER ESOPHAGUS
PNEUMOGRAPH
MYOGRAPH
SIGNAL
DISTENSION

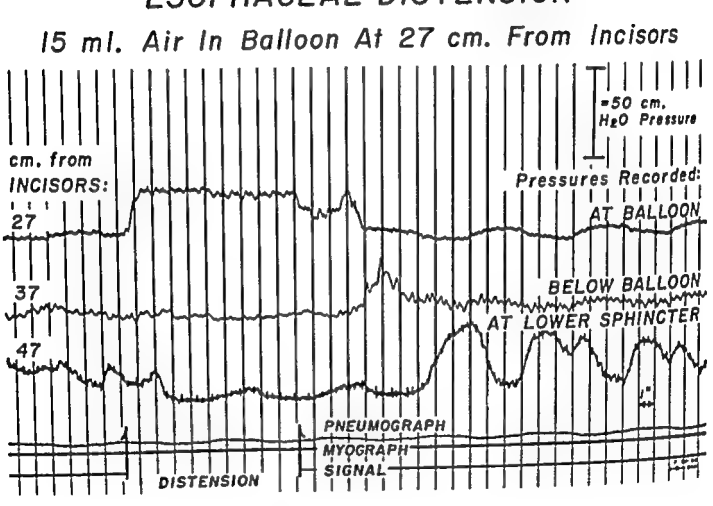


When an anchored balloon is distended in the esophagus, the response differs from that provoked by distension of the esophagus with fluid. Contractions occur above the distending balloon, but not below it.



The upper sphincter contracts vigorously during distension of the esophagus with a balloon.

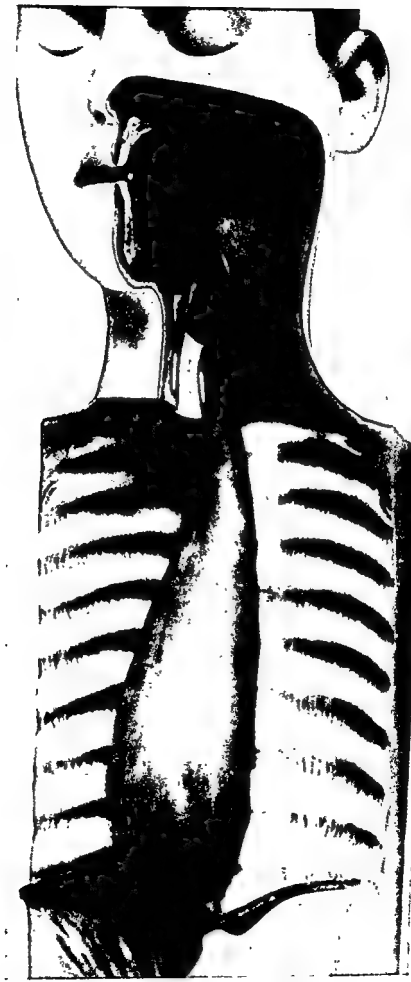
The peristaltic wave follows deflation of the balloon. The timing of its sequence is similar to that of a peristaltic wave induced by swallowing.



The gastroesophageal sphincter (lower sphincter) relaxes promptly upon distension of the esophagus with a balloon. The relaxation persists until the peristaltic wave, which follows deflation of the balloon, reaches it.

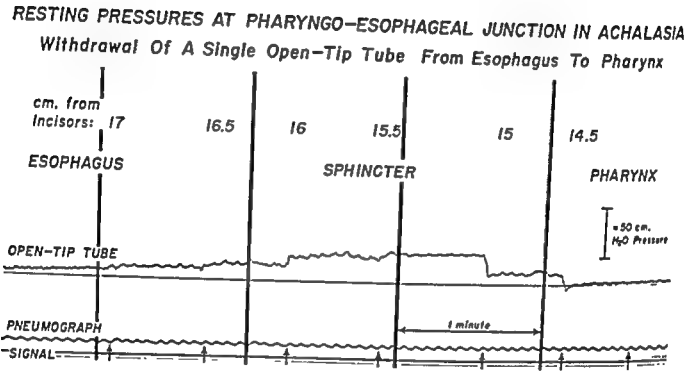
CHAPTER THREE

The Esophagus and Its Sphincters in Achalasia

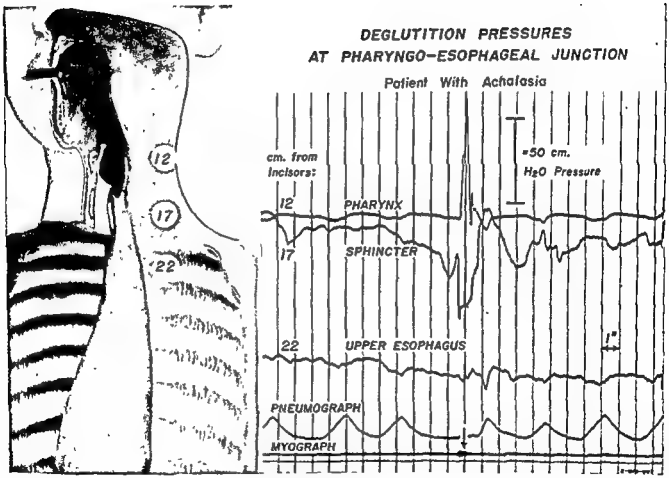


Achalasia, cardiospasm or megaesophagus is a neuromuscular disorder of the esophageal wall associated with degeneration of Auerbach's plexus. Pressures in the esophagus have been determined in more than 100 patients with this condition. A pattern of motility characteristic of the disease has emerged. The components of this pattern are normal action of the pharyngo-esophageal sphincter, absence of peristalsis in the body of the esophagus, failure of the gastroesophageal sphincter to relax with swallowing and a positive response to the administration of mecholyl.

The Pharyngo-esophageal Junction



Resting and deglutition pressures in the pharyngo-esophageal sphincter are not affected by the disease.[14]

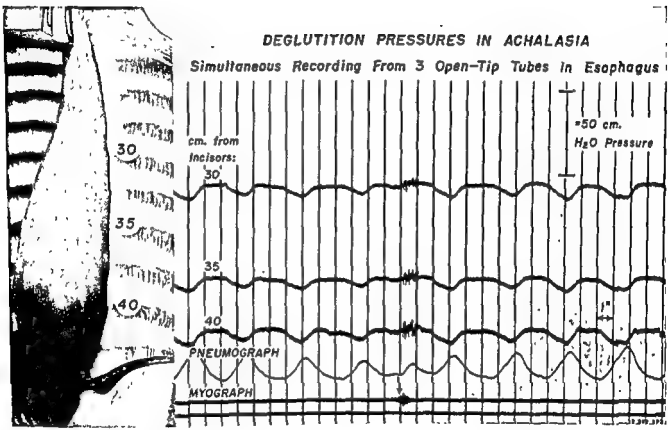

12
17
22
DEGLUTITION PRESSURES
AT PHARYNGO-ESOPHAGEAL JUNCTION
Patient With Achalasia
cm. from
Incisors:
=50 cm.
H2O Pressure
12
PHARYNX
17
SPHINCTER
22
UPPER ESOPHAGUS
1"
PNEUMOGRAPH
MYOGRAPH

The Body of the Esophagus

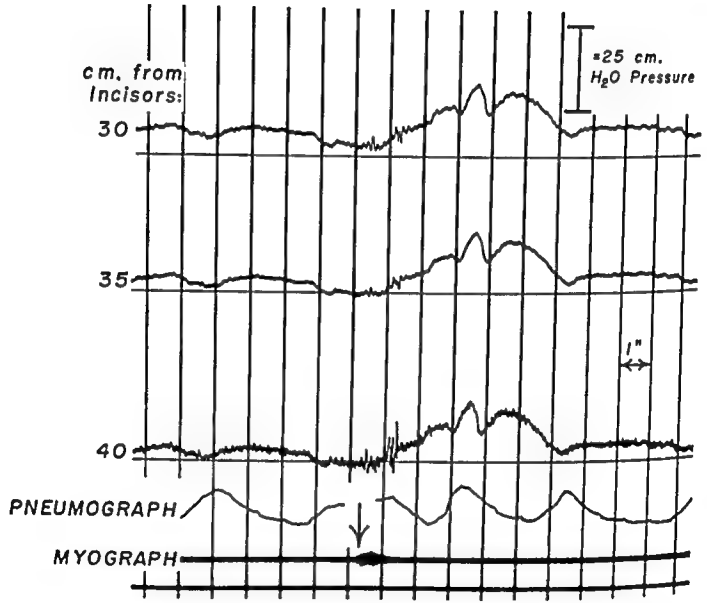
Peristalsis is absent from the esophagus of patients who have achalasia.[11, 15]

In many instances, there is no change in pressure during deglutition. This is complete motor failure of the esophagus associated with an advanced stage of the disease, and often with dilatation of the esophagus.

Deglutition

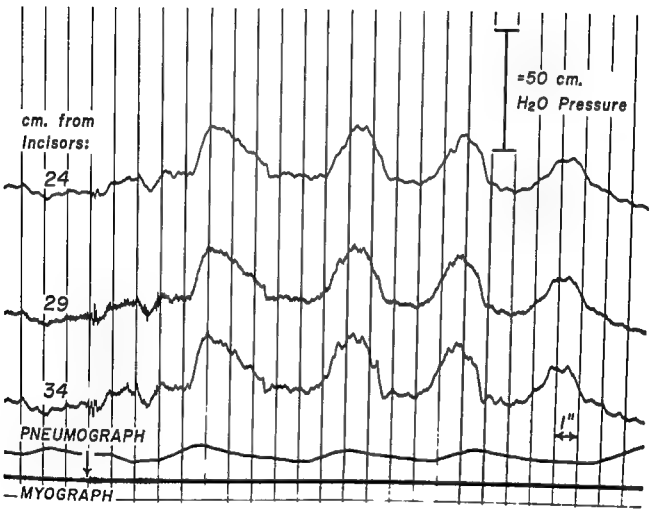


Deglutition



When an increase of pressure does occur during deglutition, the timing of the increase is abnormal. Instead of following the sequential pattern of normal peristalsis, the pressure increases simultaneously throughout the esophagus.

Deglutition

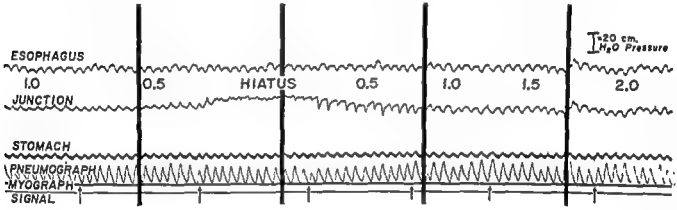


Sometimes the response to deglutition is repetitive. Even then the esophagus contracts simultaneously throughout its length. Considerable pressure may be produced in the esophagus by the contraction.

The Gastroesophageal Sphincter

RESTING PRESSURES AT GASTROESOPHAGEAL JUNCTION IN ACHALASIA

Withdrawal Of Open-Tip Tube From Stomach To Esophagus



The resting pressures at the gastroesophageal sphincter in many patients with achalasia are within normal limits.[14] In some the resting pressures are elevated.

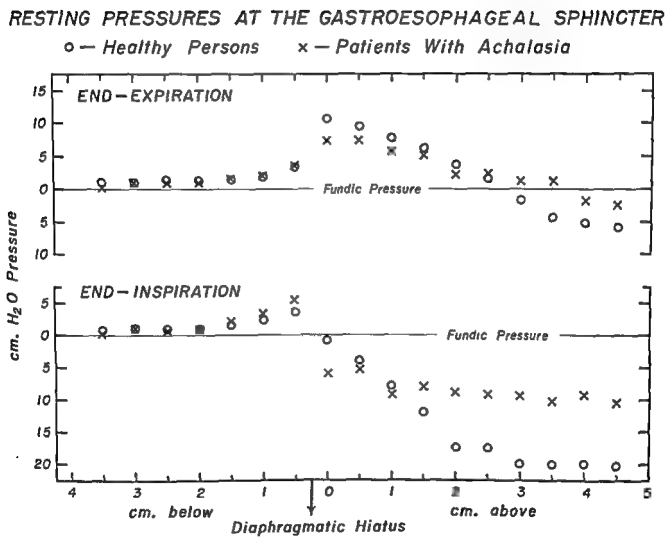
During the withdrawal of open-tip tubes from the stomach into the esophagus, a zone of elevated pressure is regularly recorded at the sphincter.

In many patients with achalasia, the resting pressures in the esophagus itself, however, are almost equal to those in the stomach.

Resting Pressures

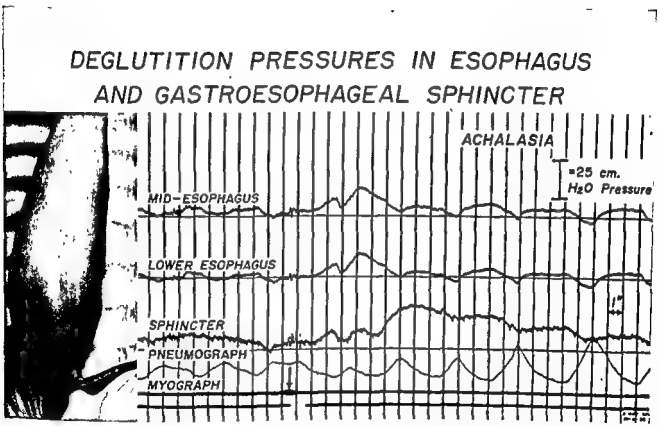
The resting pressures above and below the "hiatus" have been measured at end-expiration and end-inspiration in 13 patients with achalasia.[14] The mean values were calculated and compared with those pressures obtained by identical measurements in 16 healthy volunteers. For all practical purposes, the mean values through the sphincteric zone were identical. In none of these patients was there any indication of cardiospasm, that is, spasm of the sphincter,—but in later studies of untreated patients, some with heightened pressures at the sphincter have been encountered.

Once the sphincteric zone is passed, one difference between healthy volunteers and patients with achalasia is often evident. In achalasia, the resting pressure in the esophagus frequently is abnormally high. The high esophageal pressures are associated with dilatation of the esophagus and retention of food and secretions.


RESTING PRESSURES AT THE GASTROESOPHAGEAL SPHINCTER
o — Healthy Persons x — Patients With Achalasia
END—EXPIRATION
Fundic Pressure
END—INSPIRATION
Fundic Pressure
cm. H2O Pressure
15
10
5
0
5
10
15
20
4
3
2
1
0
1
2
3
4
5
cm. below
cm. above
Diaphragmatic Hiatus

The Gastroesophageal Sphincter Fails to Relax During Deglutition in Patients with Achalasia[16,11]

Rather often, however, this sphincter does contract. The increase in pressure which contraction of this sphincter produces is of normal amplitude and duration, but its onset is early.


DEGLUTITION PRESSURES IN ESOPHAGUS
AND GASTROESOPHAGEAL SPHINCTER
ACHALASIA
=25 cm.
H2O Pressure
MID-ESOPHAGUS
LOWER ESOPHAGUS
SPHINCTER
PNEUMOGRAPH
MYOGRAPH
1"

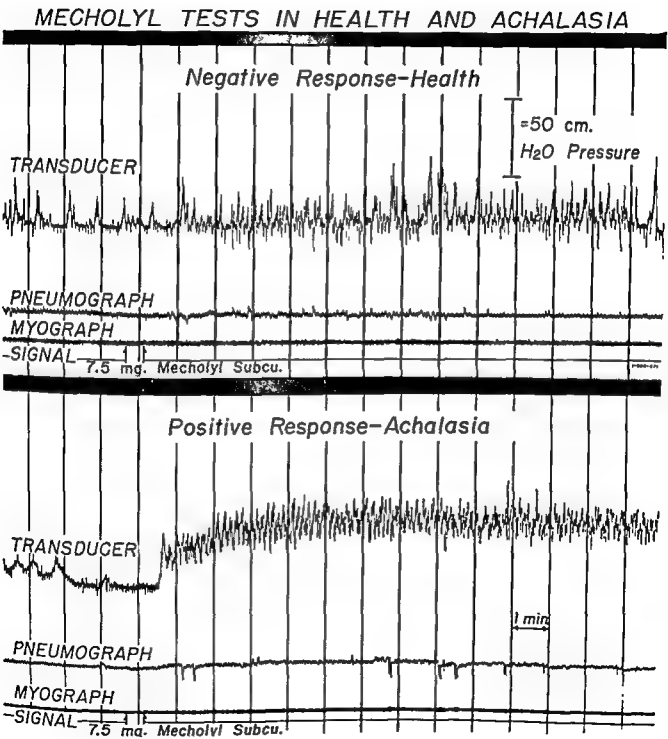
Esophagus

Response to Mecholyl

In the presence of achalasia, the esophagus is unduly sensitive to the action of mecholyl.[17, 18]

The subcutaneous injection of 5 to 10 mg. of the drug is followed by vigorous contractions of the esophagus which produce high intraluminal pressures. The esophagus may be emptied of retained products, the volume of which is sometimes as much as 500 ml. Intense substernal pain usually is associated with the increase in intraluminal pressures. Dilated esophagi may fail to respond to the drug.

Healthy persons show little or no increase in intraluminal pressure and do not experience pain.


MECHOLYL TESTS IN HEALTH AND ACHALASIA
Negative Response-Health
=50 cm.
H2O Pressure
TRANSDUCER
PNEUMOGRAPH
MYOGRAPH
SIGNAL
7.5 mg. Mecholyl Subcu.
Positive Response-Achalasia
TRANSDUCER
1 min
PNEUMOGRAPH
MYOGRAPH
SIGNAL
7.5 mg. Mecholyl Subcu.

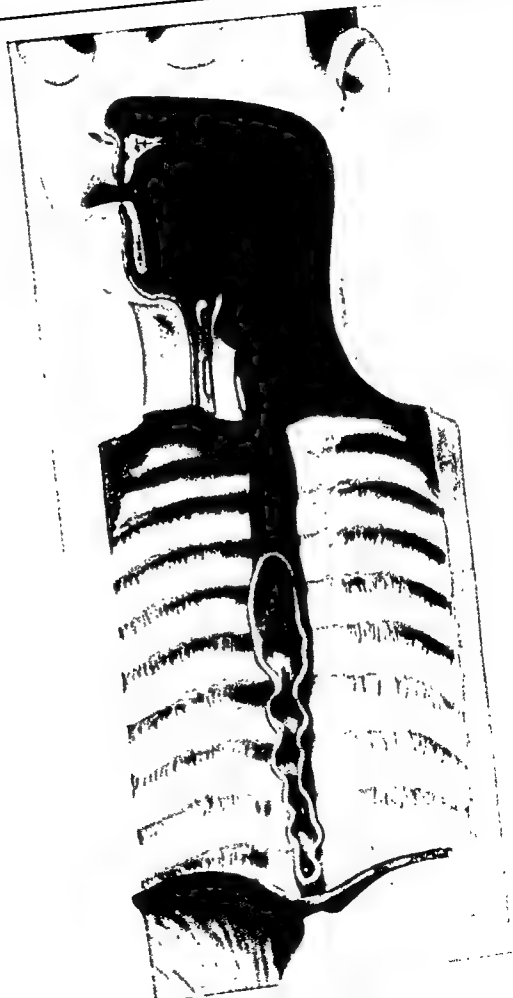
Summary

The presence of achalasia is characterized by complete loss of peristalsis in the esophagus and by failure of the gastroesophageal sphincter to relax as deglutition takes place. The obstructing factor in this disease is therefore the tone of the gastroesophageal sphincter.

Material will leave the esophagus only when the barrier offered by the tone of the sphincter is overcome by the pressure within the esophagus. Two factors are involved: the height of the column of liquid within the vertical esophagus and the pressure generated by simultaneous contraction of the entire esophagus. If these exceed the resistance of the sphincter without progressive dilatation, material will pass into the stomach and the condition may be regarded as "compensated."

CHAPTER FOUR

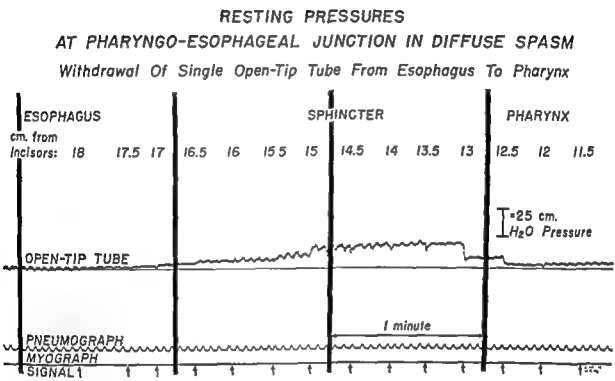
Diffuse Spasm of the Esophagus



Diffuse spasm of the esophagus is a syndrome comprised of intermittent dysphagia and substernal pain.[19-21] A characteristic abnormal pattern of motility accompanies the disorder. It may be associated with an abnormal radiologic appearance described as "curling" or "corkscrew" esophagus,[22-26] although in some cases abnormalities are not detected during a routine radiologic examination.

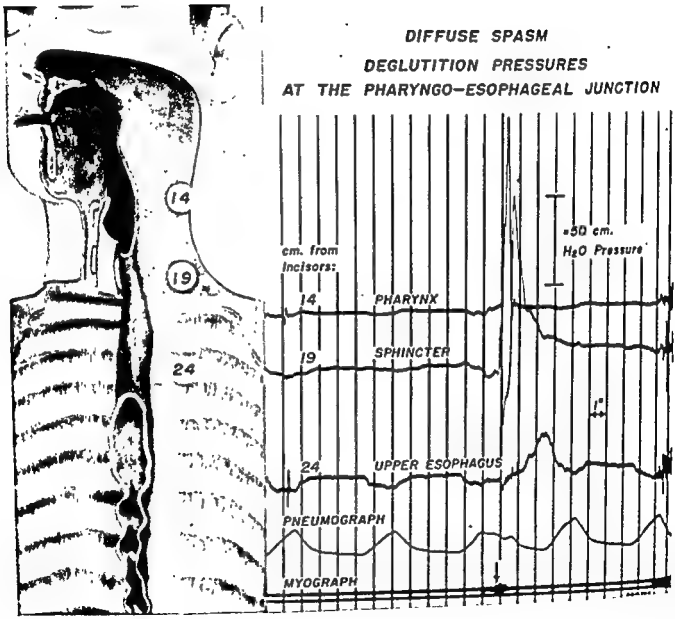
Studies of motility characterize diffuse spasm as a specific inco-ordination of the esophageal response to deglutition. Upon swallowing the peristaltic wave is obliterated or replaced in the lower part of the esophagus by a simultaneous and prolonged contraction of this region. The contraction generally is premature, repetitive and produces abnormally high pressures.[27]

The Pharyngo-esophageal Sphincter



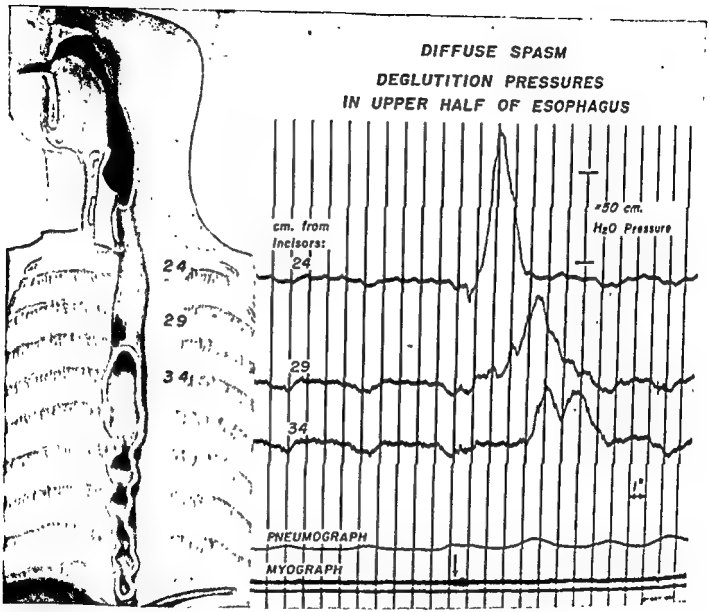
Resting pressures through the pharyngo-esophageal junction are within normal limits in patients with diffuse spasm.

Deglutition



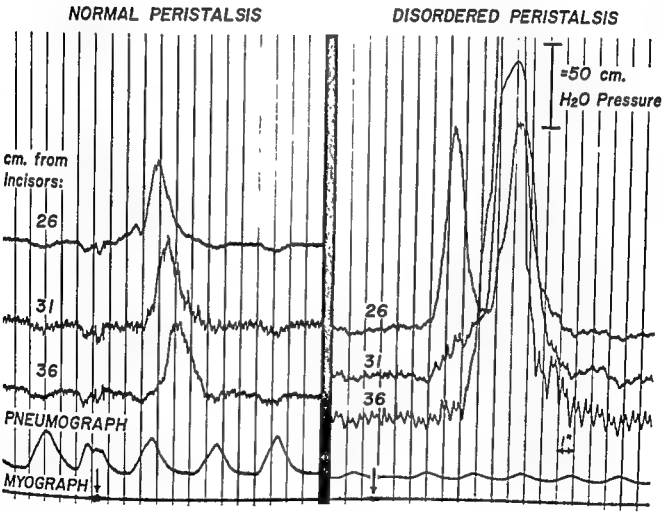
As deglutition takes place, the pharyngo-esophageal sphincter relaxes and then contracts as it does in health, and a normal peristaltic wave usually is initiated in the upper part of the esophagus.

The Body of the Esophagus



However, even in the upper part of the esophagus, some abnormality may be present. Most commonly this is a double response to deglutition (34 cm. from the incisors).

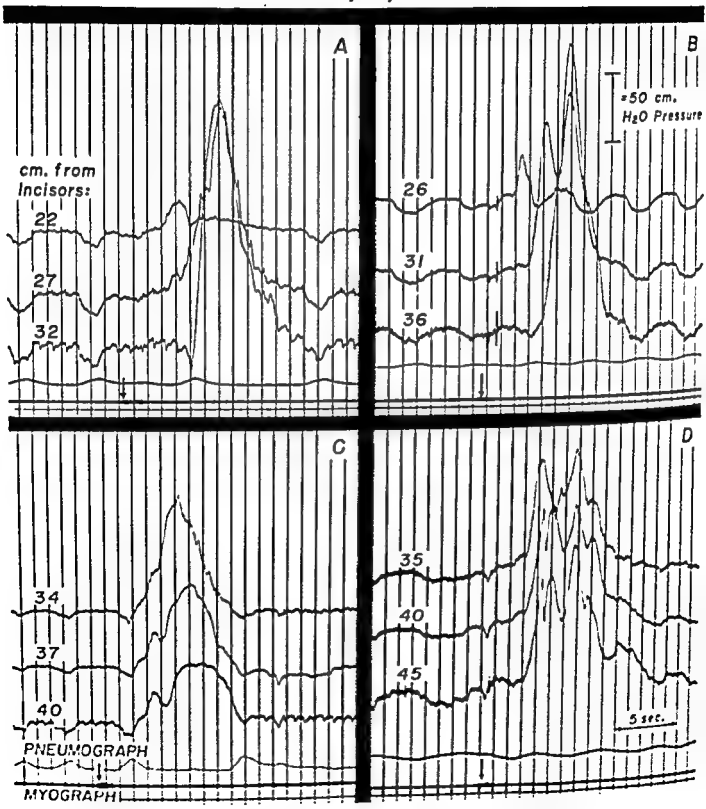
CONTRAST BETWEEN DEGLUTITION RESPONSE IN HEALTH AND IN DIFFUSE SPASM



In the lower half of the esophagus the normal peristaltic wave is obliterated by an abnormal contraction. Characteristically, this is a simultaneous contraction of the entire lower portion of the esophagus. This simultaneous contraction occurs earlier in the deglutition complex than would a normal peristaltic wave.

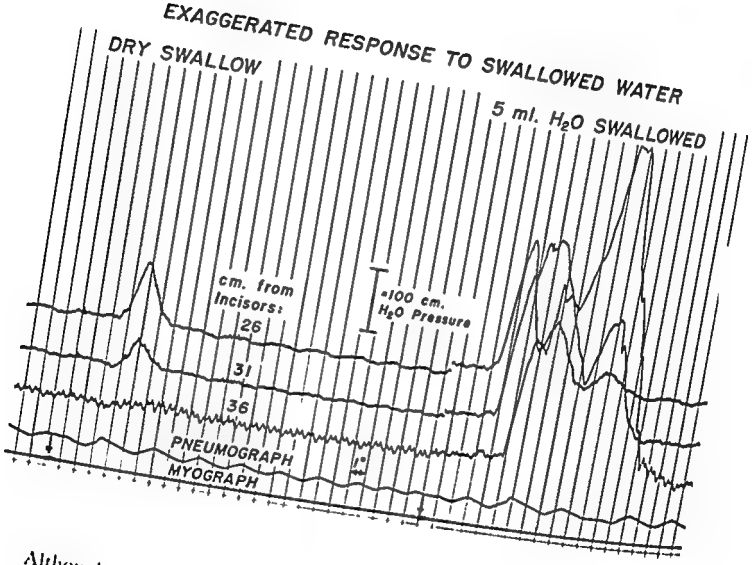
DEGLUTITION PRESSURES IN DIFFUSE SPASM

Patients A, B, C and D

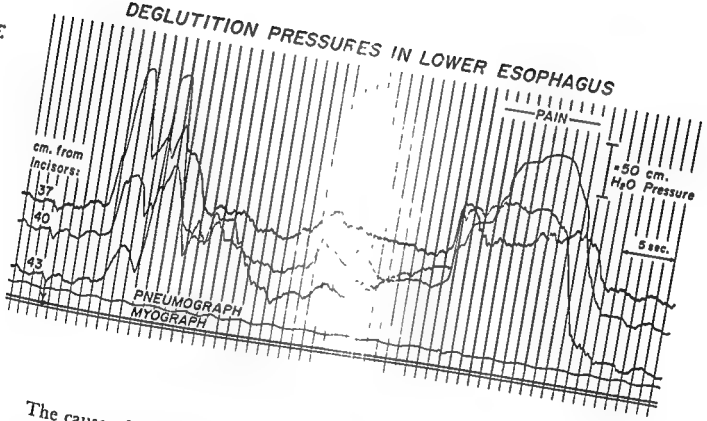


Deglutition

Generally, the abnormal contractions are prolonged and often repetitive. Two or more pressure peaks may follow a single swallow. The pressure developed usually is extremely high, often exceeding 200 cm. of water.

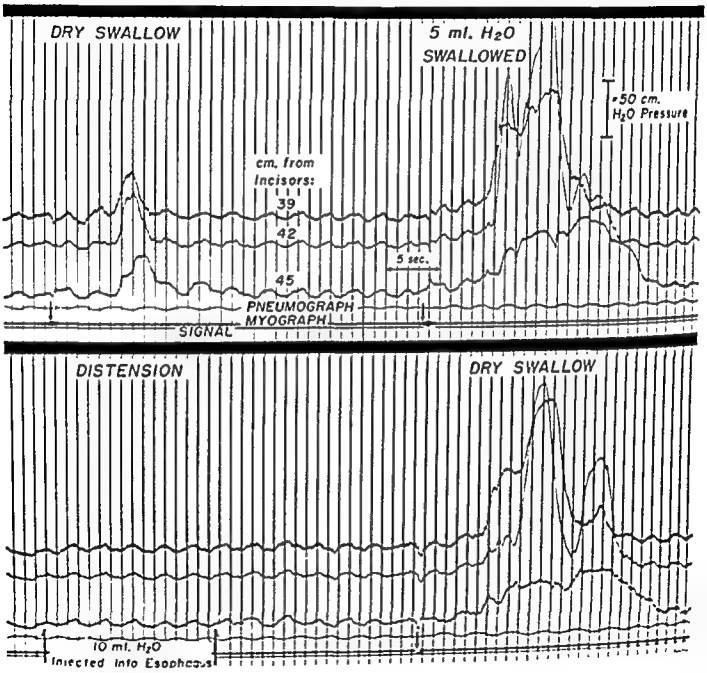


Although the abnormalities of the deglutition response customarily accompany "dry" or "empty" swallows, as a rule they are greatly exaggerated when water or solid material is swallowed. In particular, the pressure caused by the contractions is increased, and values as high as 500 cm. of water have been recorded.



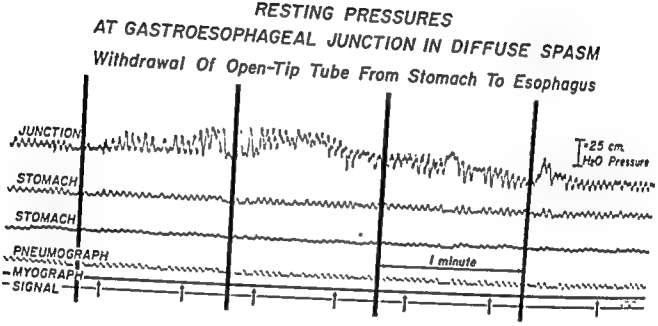
The cause of the substernal pain of patients who have diffuse spasm has not been definitely established. Exceedingly high pressures of 500 cm. of water, and more, have been recorded in such patients without complaints of pain. However, in some patients during a spontaneous attack of substernal pain, a prolonged elevation of the intraluminal pressures has been recorded in the lower 6 cm. of the esophagus. Relief has accompanied a return of the esophageal pressure to normal limits.

EXAGGERATED DEGLUTITION RESPONSES IN THE PRESENCE OF DISTENSION

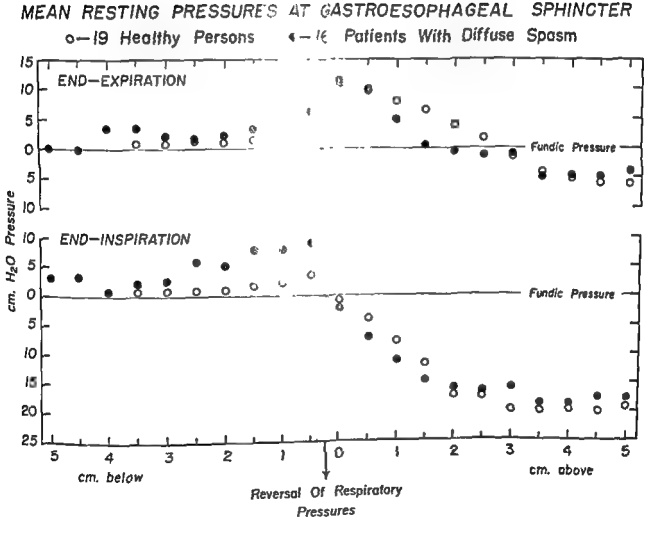


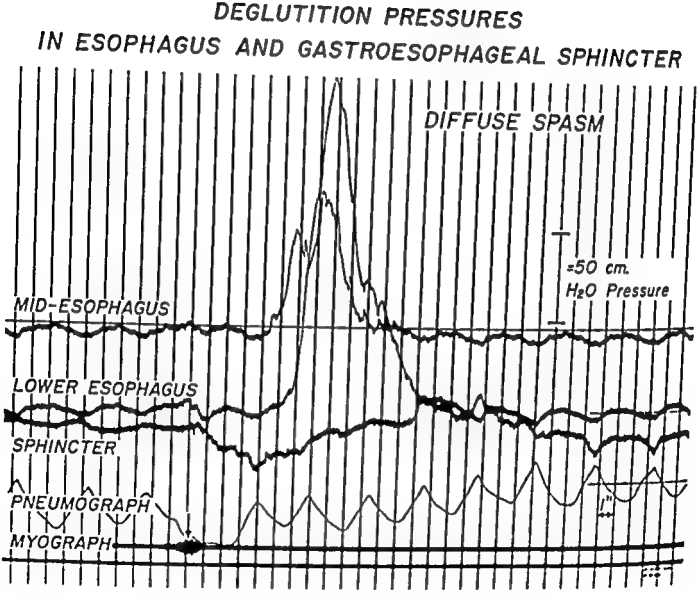
The exaggerated response is due to the distension of the esophagus by the water.

The Gastroesophageal Sphincter



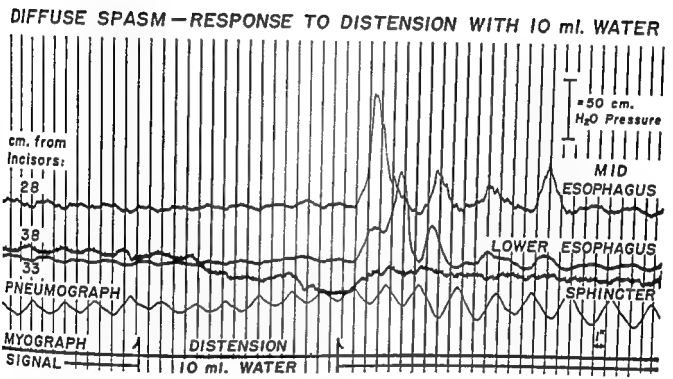
Withdrawal of the pressure-sensitive units through the gastroesophageal junction generally discloses a broader band of elevated pressure than is encountered in health. The pressure also is higher. The extra width of the zone generally is distal to the point of respiratory reversal.


MEAN RESTING PRESSURES AT GASTROESOPHAGEAL SPHINCTER
o—19 Healthy Persons
•—16 Patients With Diffuse Spasm
END—EXPIRATION
END—INSPIRATION
Fundic Pressure
Fundic Pressure
cm. H2O Pressure
15
10
5
0
5
10
15
20
25
5 4 3 2 1 0 1 2 3 4 5
cm. below
cm. above
Reversal Of Respiratory Pressures



As swallowing takes place, the gastroesophageal sphincter relaxes and then contracts in a normal manner.

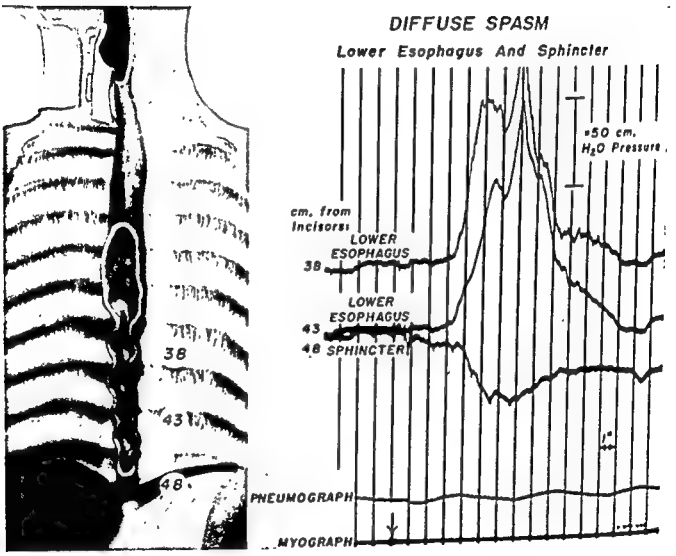
Distension



Even in the absence of swallowing, prompt relaxation of the sphincter accompanies distension of the upper part of the esophagus. Later, abnormal repetitive contractions may occur in the mid and lower parts of the esophagus.

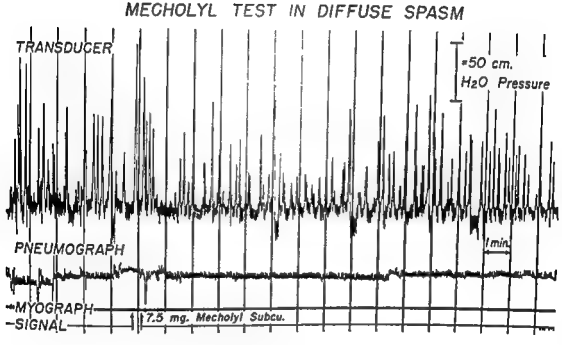
Thus, in diffuse spasm, the gastroesophageal sphincter responds normally to deglutition and to distension. The sphincter offers no obstruction to the passage of the esophageal content.

Deglutition



Although relaxation of the sphincter occurs, its effect may be negated during swallowing by premature and prolonged contractions of the lower part of the esophagus. Thus, the obstruction in the presence of diffuse spasm is not at the level of the sphincter, but in the lower part of the esophagus.

The Esophageal Response to Mecholyl



The response to the injection of methacholine chloride (mecholyl) is rarely positive in patients who have diffuse spasm. If an increase in pressure is recorded, it is localized to the lower part of the esophagus and is not generalized, as it is when achalasia is present.

Summary

Diffuse spasm is characterized by inco-ordination of the response of the lower part of the esophagus to deglutition and esophageal distension. Simultaneous and prolonged contraction of the lower part of the esophagus is the salient physiologic finding. The radiologic appearance of curling of the esophagus is the result of the forcing of barium into zones of lesser resistance or strength by the vigorous contraction of the whole affected area. There is no physiologic evidence of alternating rings or segments of contraction and relaxation.

The extreme pressures developed by the contractions have not been recorded in the presence of health. They are most likely the consequences of the muscular hypertrophy which has been demonstrated in this condition.[28] This hypertrophy may, in turn, be produced by the increased work of the repetitive and prolonged contractions. It is not surprising that permanent diverticula may develop. It also seems likely that the shortening of the esophagus which must accompany forceful and prolonged contraction of longitudinal muscle fibers produces hiatal herniation.

Summary

Diffuse spasm is characterized by inco-ordination of the response of the lower part of the esophagus to deglutition and esophageal distension. Simultaneous and prolonged contraction of the lower part of the esophagus is the salient physiologic finding. The radiologic appearance of curling of the esophagus is the result of the forcing of barium into zones of lesser resistance or strength by the vigorous contraction of the whole affected area. There is no physiologic evidence of alternating rings or segments of contraction and relaxation.

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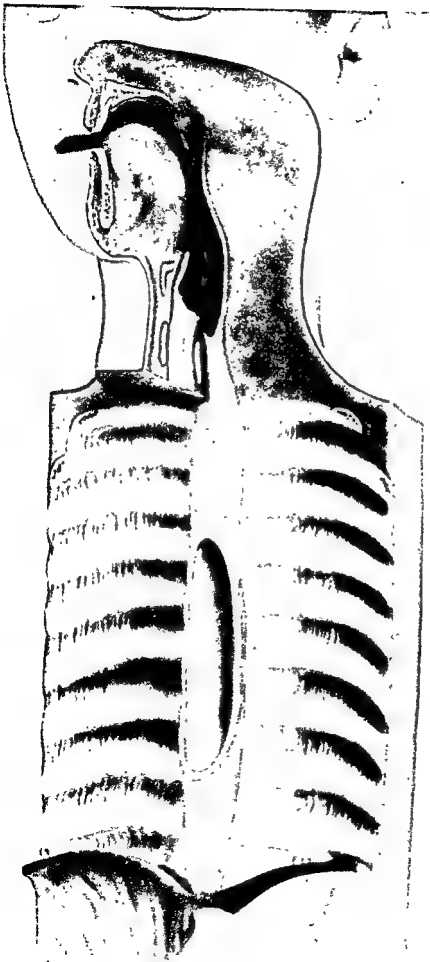
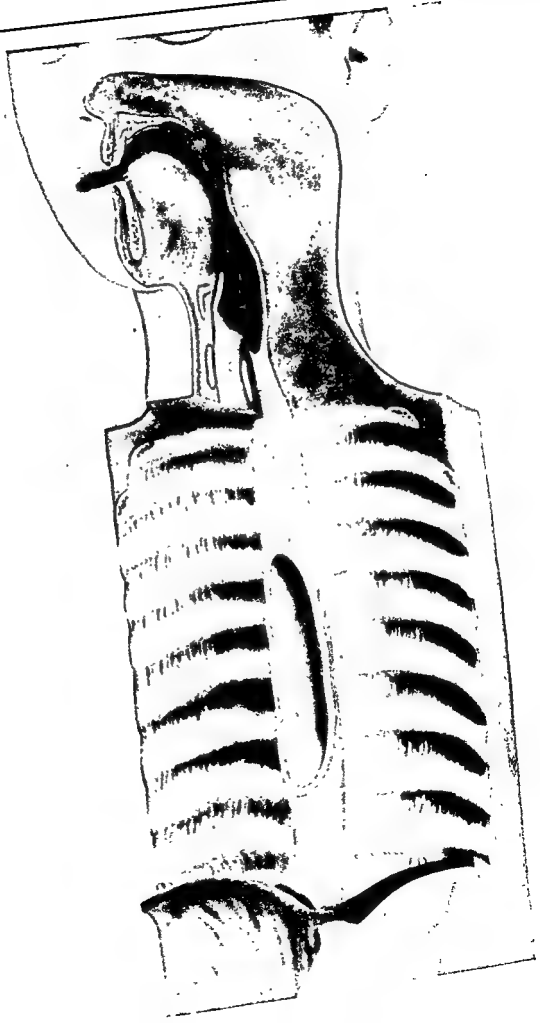
CHAPTER FIVE

Sclerodermal Involvement of the Esophagus *and Its Sphincters*



The esophagus often is involved when scleroderma is present. The mucosa of the esophagus is thickened and the musculature is degenerated. The patients may complain of dysphagia, heartburn and retrosternal pain. Reflux esophagitis, formation of stricture, shortening of the esophagus and hiatal hernia sometimes are found. Studies of motility have revealed the physiologic mechanism of the changes.



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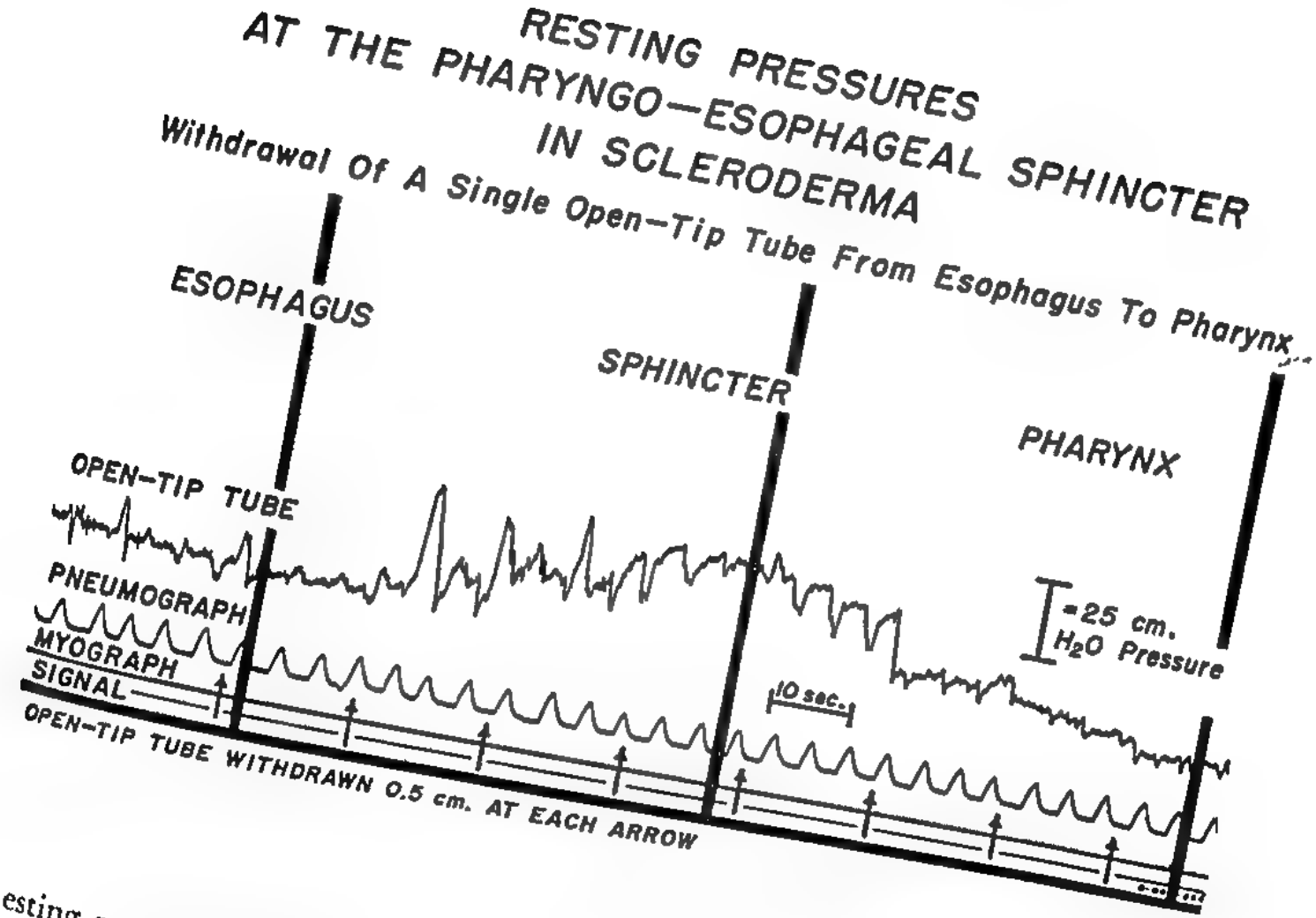
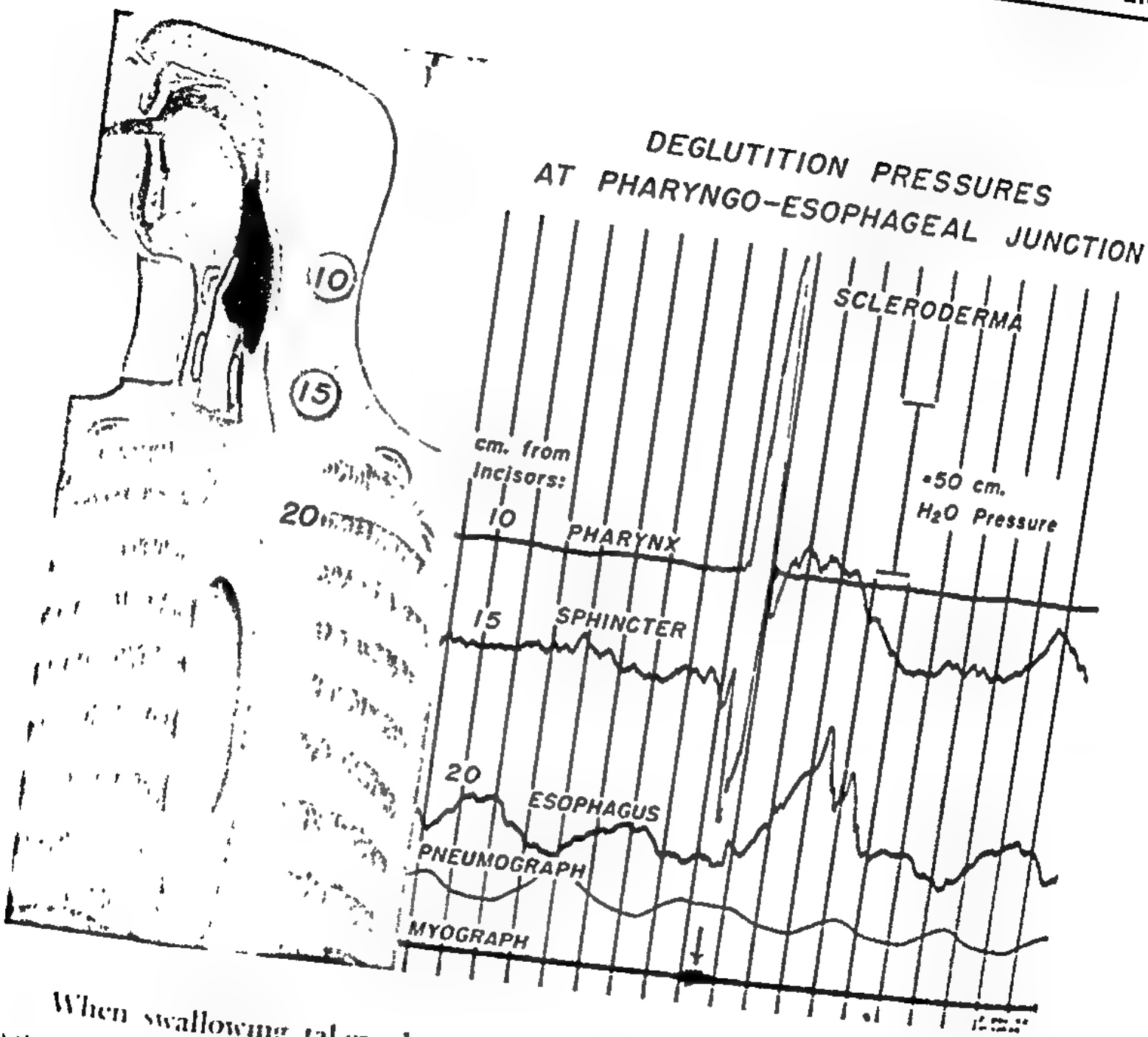
The Pharyngo-esophageal Sphincter



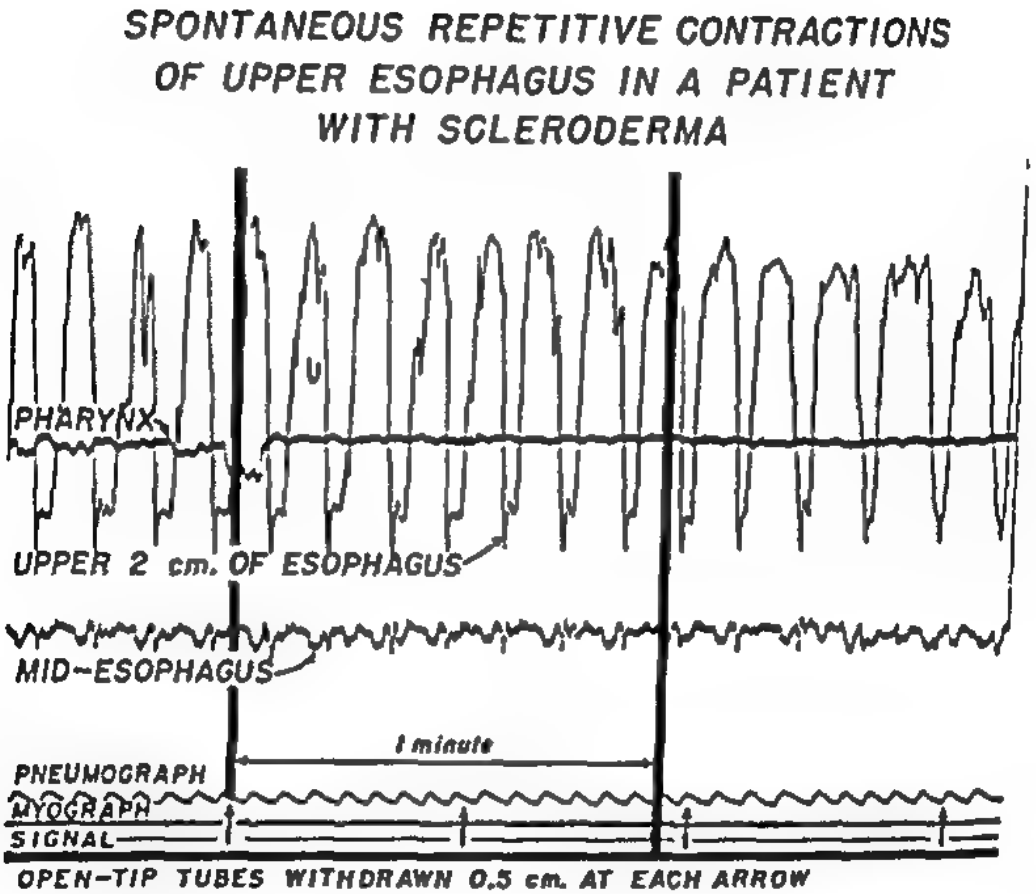
Resting pressures at the pharyngo-esophageal sphincter in patients with sclero-
derma are within the range found in the presence of health.



When swallowing takes place, the pressure in the sphincter decreases as that in the pharynx increases. Thus, the sphincter relaxes as the pharynx contracts. Then the pressure in the sphincter increases as the peristaltic contraction descends through the sphincter into the upper part of the esophagus. The temporal sequence and the magnitude of the changes in pressure are normal.

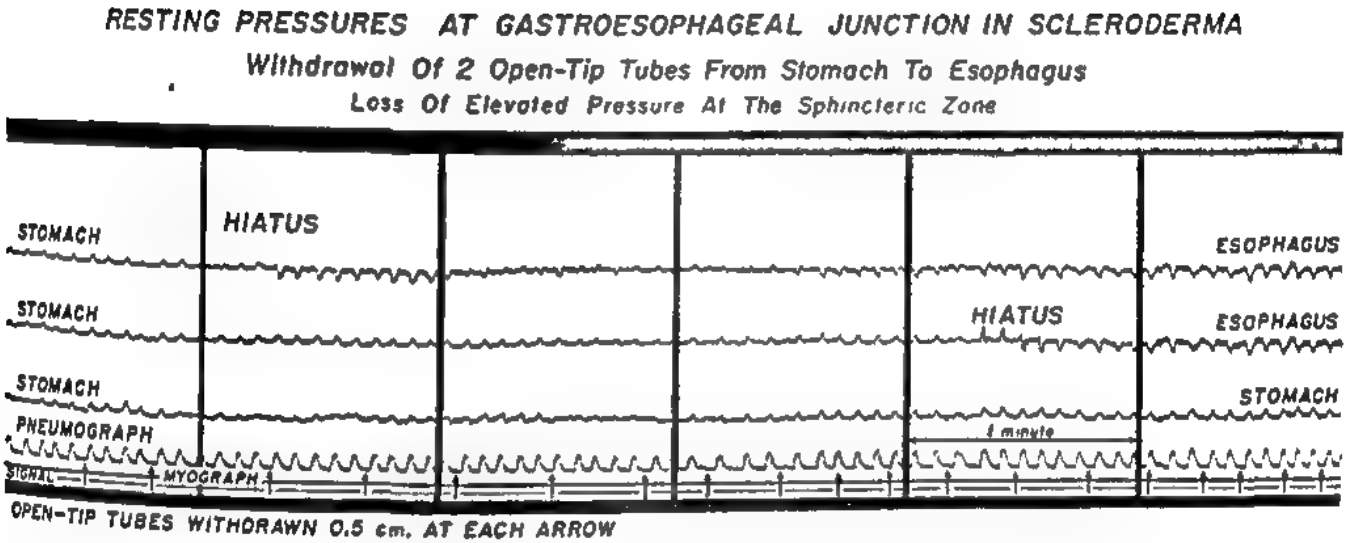
The Body of the Esophagus

Resting Pressures



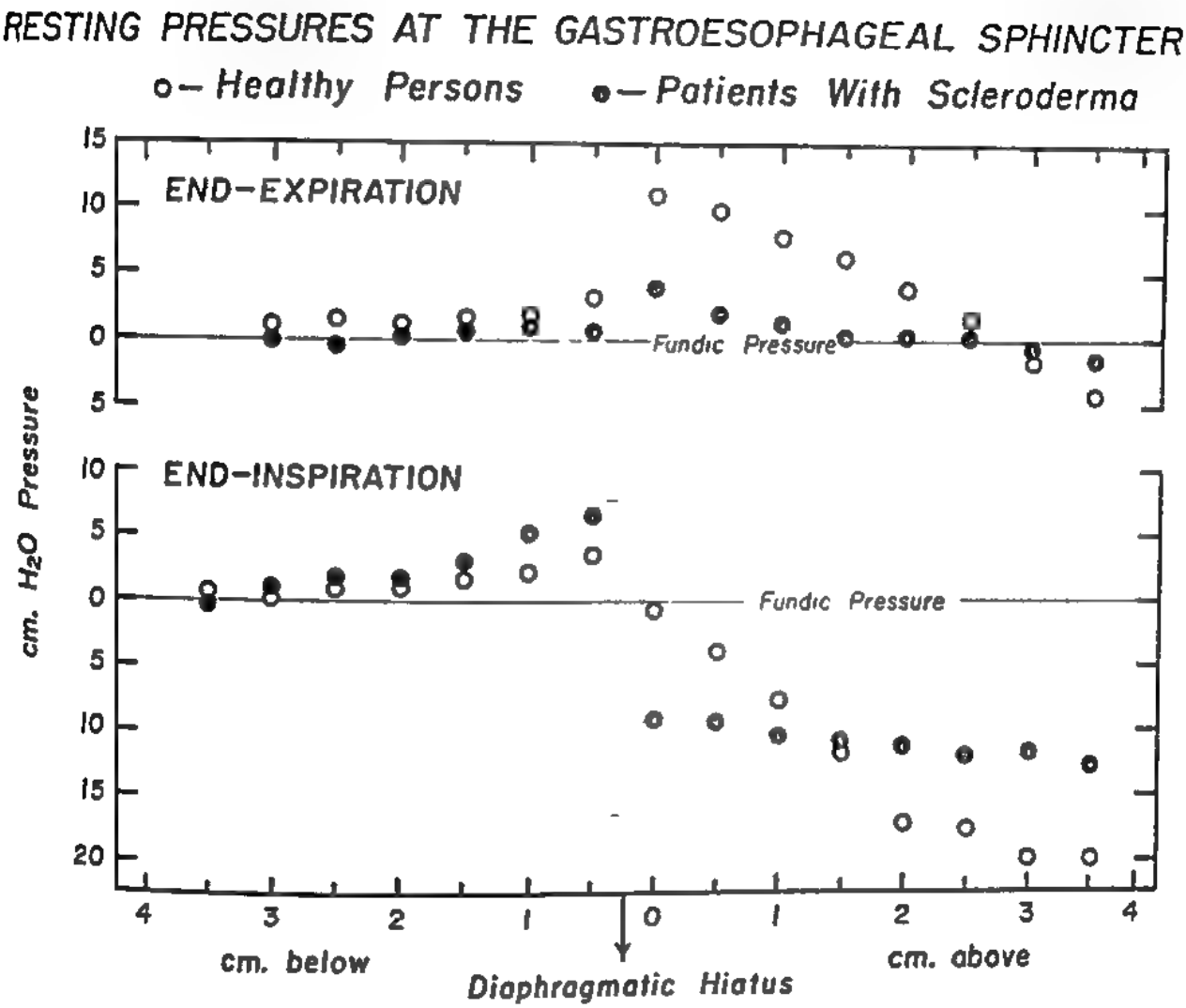
Generally, the resting pressures in the body of the esophagus are higher than those found in the presence of health. Spontaneous contractions sometimes are recorded in the upper few centimeters of the esophagus. Occasionally, these are a marked, although localized, feature.

Resting Pressures



In advanced stages of scleroderma there is no evidence of a zone of increased pressure between the stomach and the esophagus.[7] The pressures in the two organs are equal, and gastric contents may reflux.[29, 30]

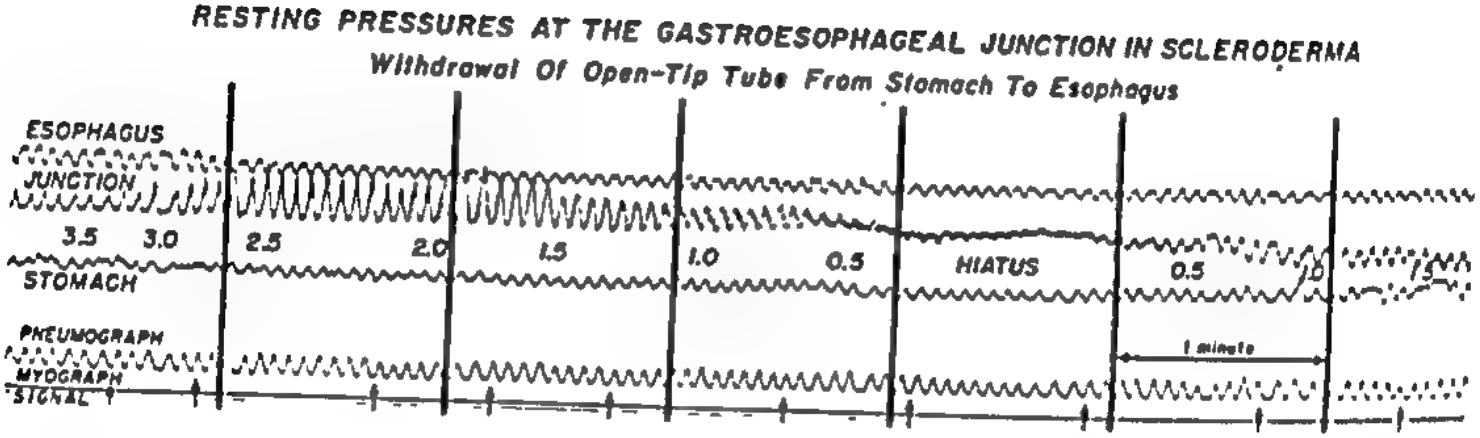
The Gastroesophageal Junction



When the mean resting pressures at the gastroesophageal junction of nine patients with scleroderma were determined and compared with those in 16 healthy persons, the end-inspiratory pressures below the "hiatus" were clearly higher in the patients with scleroderma, while the end-expiratory pressures were definitely lower.[7]

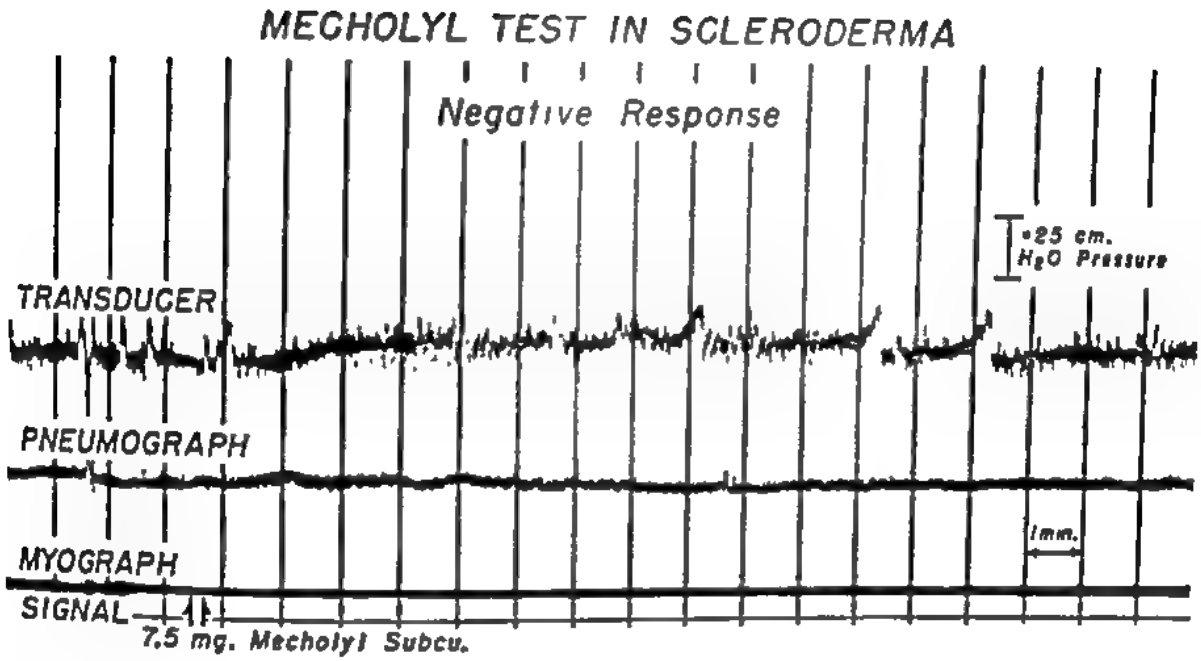
Thus, there is a partial failure of the gastroesophageal sphincter in most patients who have scleroderma and a complete failure in some.

Resting Pressures

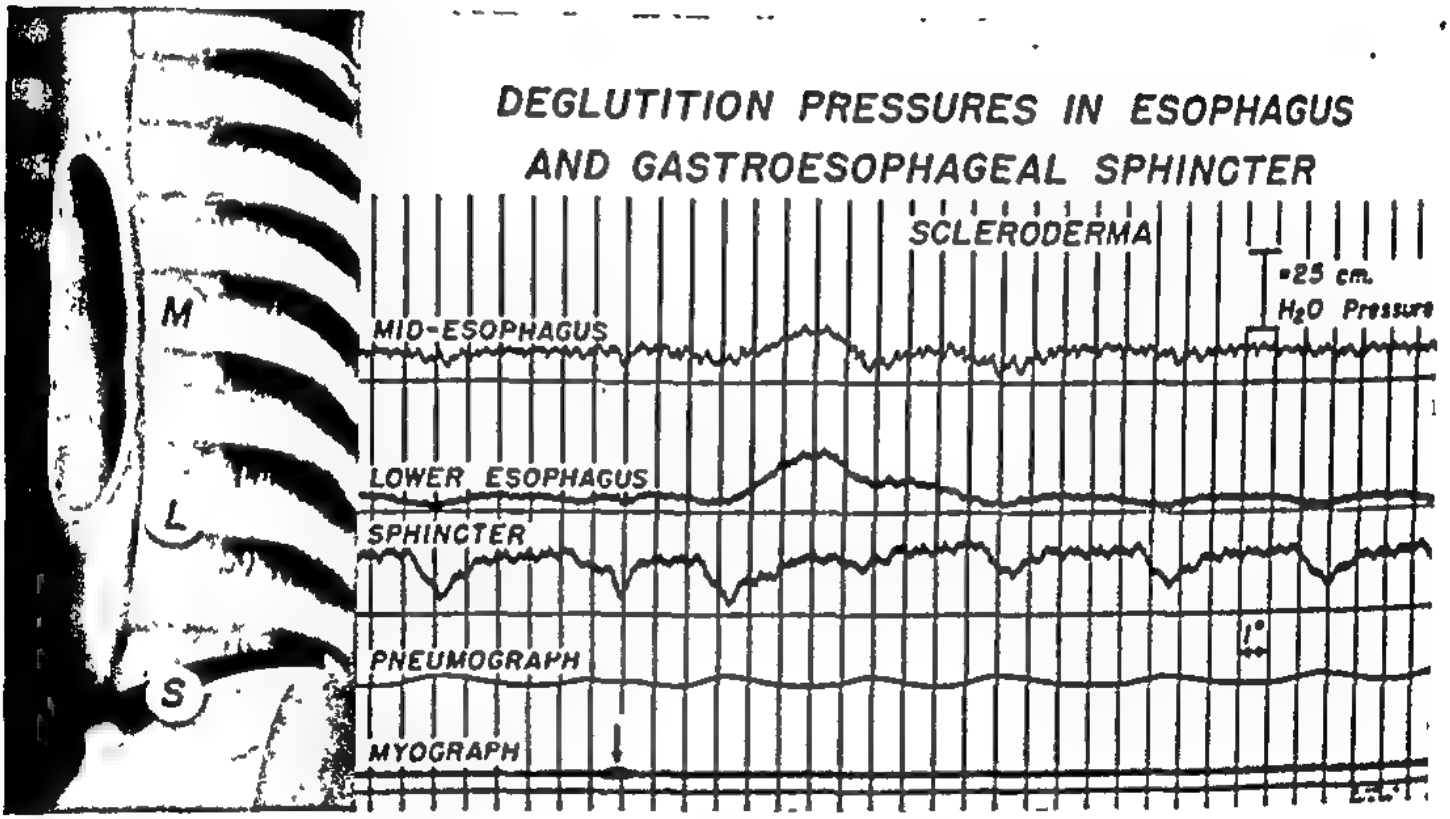


In most patients with this disease, as open-tip tubes are withdrawn through the gastroesophageal junction, a marked increase of end-inspiratory pressure is recorded as the "hiatus" is approached, but there is little change in end-expiratory pressure anywhere in the zone.

Response to Mecholyl

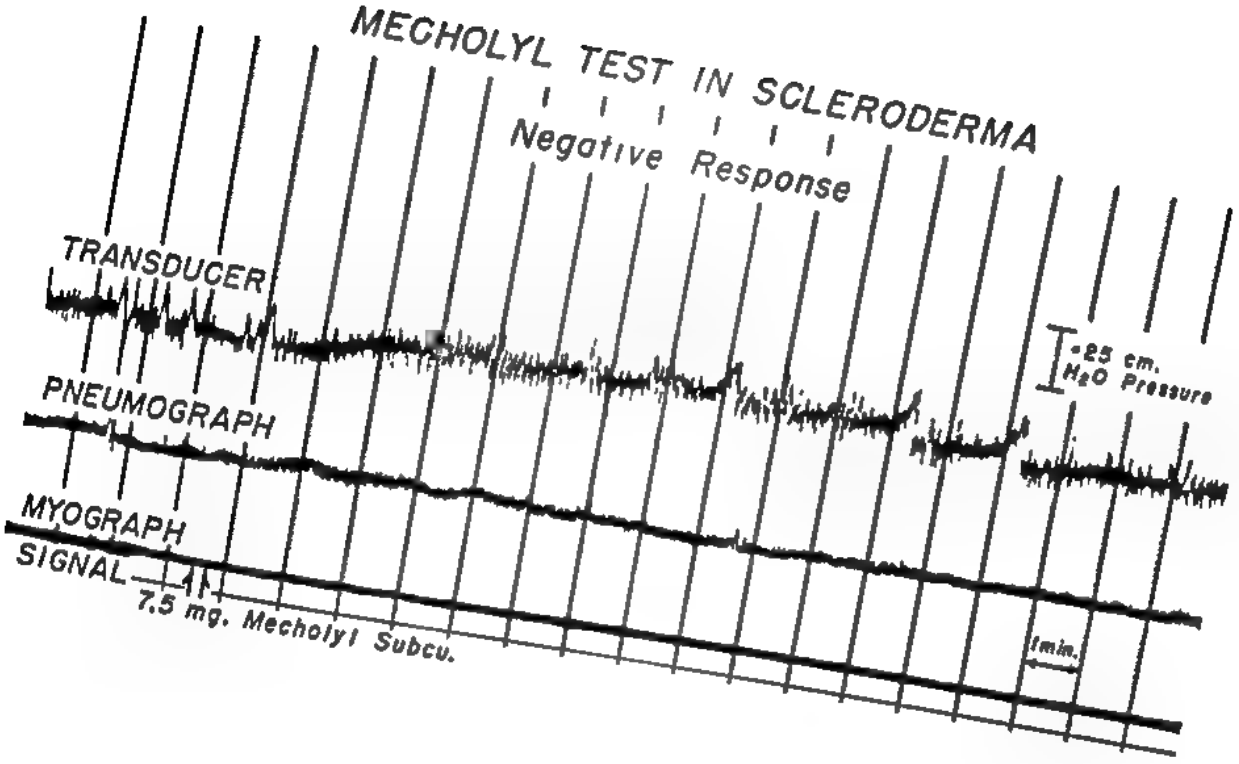


The response to the administration of mecholyl in the presence of scleroderma is negative.



In most patients with scleroderma, no decrease in pressure occurs at the gastroesophageal sphincter when swallowing takes place.

Response to Mecholyl



e response to the administration of mecholyl in the presence of scleroderma is negative.

Summary

The two abnormal features of esophageal motility in the presence of scleroderma which predispose to the development of symptoms and clinical findings are the loss of tone of the gastroesophageal sphincter and the loss of contractile power in the lower three quarters of the esophagus. The loss of tone in the sphincter allows regurgitation of gastric contents, and the loss of contractile power in the lower part of the esophagus renders that portion of the organ powerless to react and thus to expel the foreign material. Esophagitis, shortening and formation of stricture may follow.

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INDEX